Send us your lame... your chair-bound too!

RECREATION CENTER *for* THE HANDICAPPED INC.

207 Skyline Boulevard • 665-4100
San Francisco, California 94132

3/5/75.

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CONTRIBUTIONS TO SUPPORT THIS VITALLY NEEDED WORK ARE TAX DEDUCTIBLE

6/12/80

THE CHILDCARE/ STUDY CENTER.

H.E. Thelander M.D.

A pleasant addition to Home Coming day was a visit to the Daycare Study/ Center. During the years I spent at UCSF I saw the need for such a facility, and spent almost a year studying the day care centers and nursery schools of San Francisco, making that the topic for my" Senior " arbeit. I have visited similar centers in Russia, European Countries and in Israel. United States has been slow in recognizing this need;- the general opinion has been " mothers should stay home and take care of their babies". I am not adverse to that idea either but babies and young children need more than apron strings. In the nursery school for handicapped children at Childrens Hospital,mother sometimes wept because the child was reluctant to leave when picked up to go home. The same thing was said by teachers in other nursery schools. The opportunity to be with others of the same age in a facility planned for their size and equipped and conducted for their age is a discovery for them.

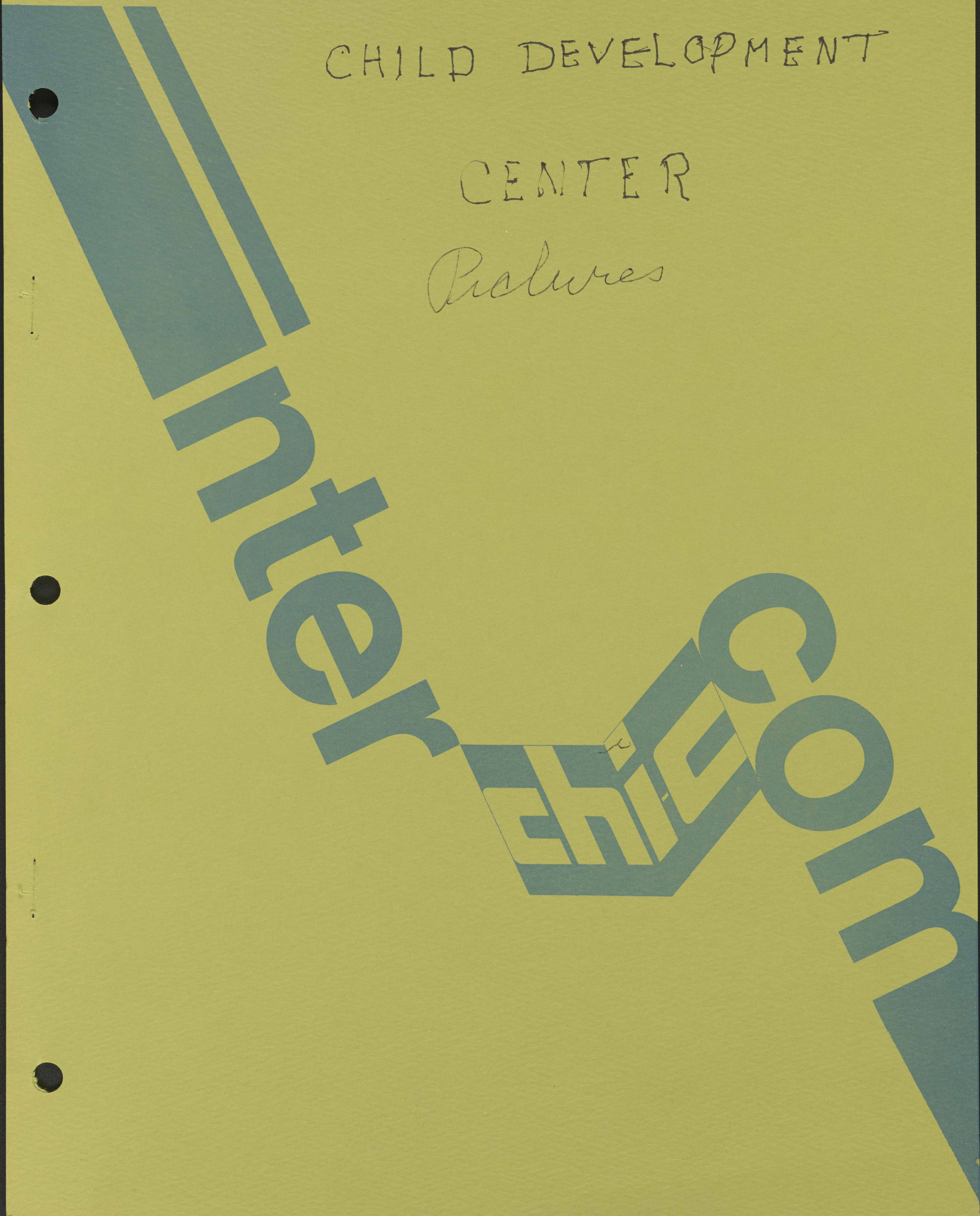
The center, properly, includes the name "Study" in its title. Medical and other health students should study growth and development as an essential part of their preparation, physical, mental and emotional. Medical schools and physicians are now constantly criticized for devoting all their time to research and disease. To some one who practiced during the years say from mid twenties to mid sixties, this is understandable. When the entire day and often late into the night, the time was devoted to diseases such as diphtheria, smallpox, meningitis, poliomyelitis and other highly fatal diseases with the necessity too often of telling parents, " This is fatal. We have no cure"., It is easy to see why doctors wanted and demanded research to find cures and prevention. This could only be done by investigation in laboratories.

Times have changed. We are in a social revolution. We must learn to live together with all Nationalities and all colors. We must learn to understand human growth and development in all areas, physical , mental and emotional. We need to study the prevention of social diseases as well as physical and mental ones. Crimes are rampant,- murder, arson, rape and many others. Lets study human development!

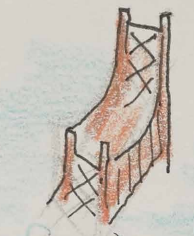
CHILD DEVELOPMENT

CENTER

Pictures



SLIDE I



SAN FRANCISCO STATISTICS

POPULATION — — — — — 750,000

CHILDREN UNDER 20 YEARS - - - 199,000

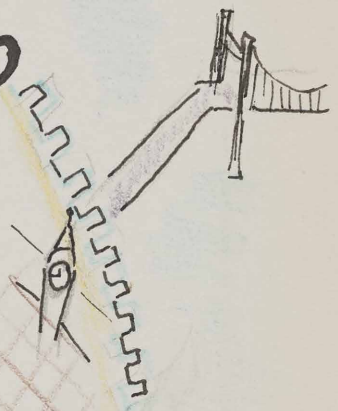
CHILDREN ON ANC - - - 12— 15,000

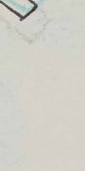
SEVERELY HANDICAPPED - - - - 2,000

BLIND, DEAF
HALT AND RETARTED

FRINGE CASES — — — — 12-20% OF ALL CHILDREN

EMOTIONALLY DISTURBED
DEPRIVED
NEUROLOGICALLY HANDICAPPED
WITH LEARNING PROBLEMS
ANOMALIES AND OTHER PHYSICAL HANDICAPS
BORDERLINE IQS.





CHILD DEVELOPMENT CENTER

(FOR DIAGNOSIS, TOTAL ASSESSMENT FOLLOW THROUGH AND COUNSELING.

STAFFS:

MEDICAL: PEDIATRIC EVALUATION AND SPECIALTY CONSULTATIONS.

ANCILLARY:

PSYCHOLOGIST
EDUCATOR
SPEECH-AUDIOLOGIST
READING ANALYST
SOCIAL SERVICE
PHYSIOTHERAPIST
OCCUPATIONAL THERAPIST

SECRETARIAL:

VOLUNTEERS:

REDEVELOPMENT PLANS

I. THERE SHOULD BE NO INDIGENT CHILDREN AND NO DISCRIMINATION BECAUSE PARENTS CANNOT PAY. FEDERAL. STATE AND LOCAL PUBLIC HEALTH AGENCIES SHOULD CONTRACT FOR CARE AT PRIVATE OR UNIVERSITY HOSPITALS.

II A DIAGNOSTIC AND ASSESSMENT FACILITY AS DESCRIBED UNDER CHILD DEVELOPMENT SHOULD BE AS ESSENTIAL A PART OF A PEDIATRIC DEPARTMENT AS THE CLINICAL LABORATORIES ARE WITH A PATHOLOGIST IN CHARGE OF A PARAMEDICAL STAFF. IT SHOULD BE AVAILABLE TO ALL PHYSICIANS FOR REFFRRAL OF PATIENTS NOT FUNCTIONING NORMALLY.

SLIDE V

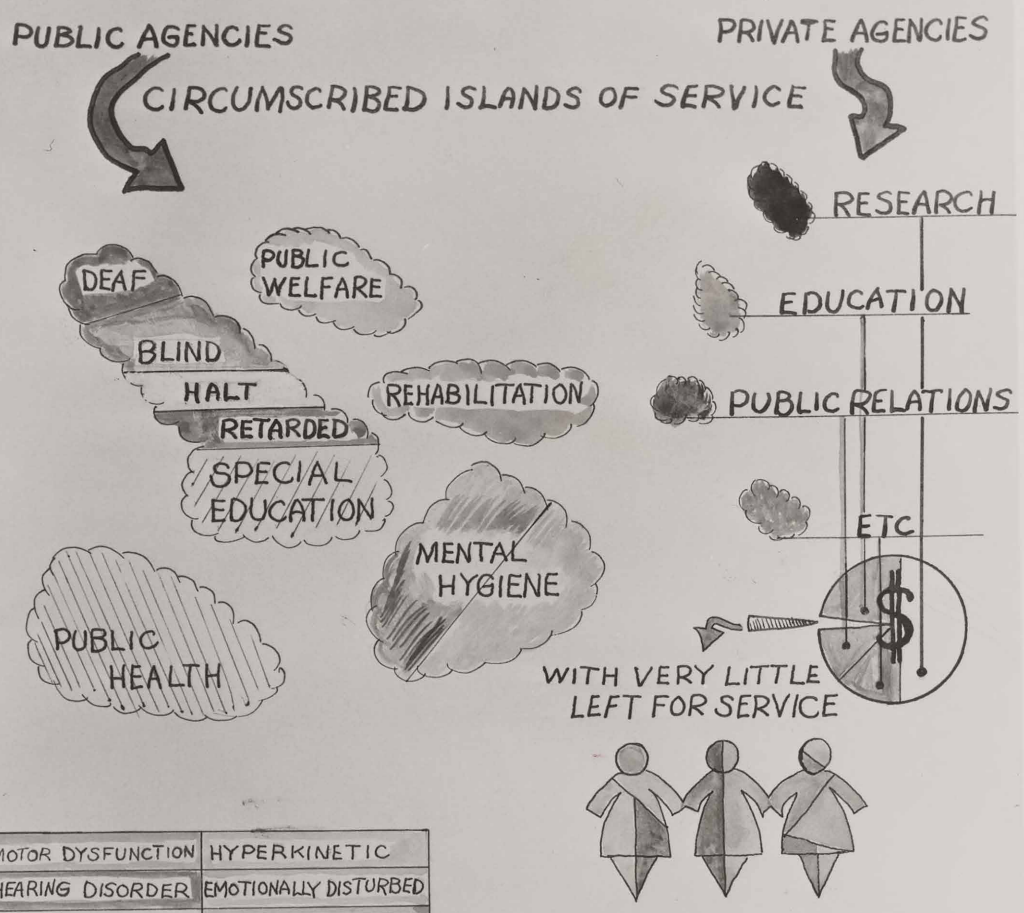
REDEVELOPMENT CONTINUED

III THE NUMBER AND TYPES OF PROBLEMS ASSOCIATED WITH OUR AGING POPULATION CALLS FOR MORE HOSPITALS EXCLUSIVELY FOR ADULTS.

CHILDRENS PROGRAMS SHOULD BE DEVELOPED IN CLINICAL CENTERS FOR CHILDREN.

THE HABITUATION OF HOSPITAL BOARDS, ADMINISTRATIONS, STAFFS, AND ACCREDITATION BOARDS TO THINK IN TERMS OF FOUR CLINICAL DEPARTMENTS SURGERY, OBSTETRICS, MEDICINE. PEDIATRICS, FRAGMENTS THE CHILDRENS SERVICES MAKING IT IMPOSSIBLE TO HAVE ADEQUATE DEPARTMENTS FOR PROPER PROGRAMING OR TEACHING.

SLIDE III

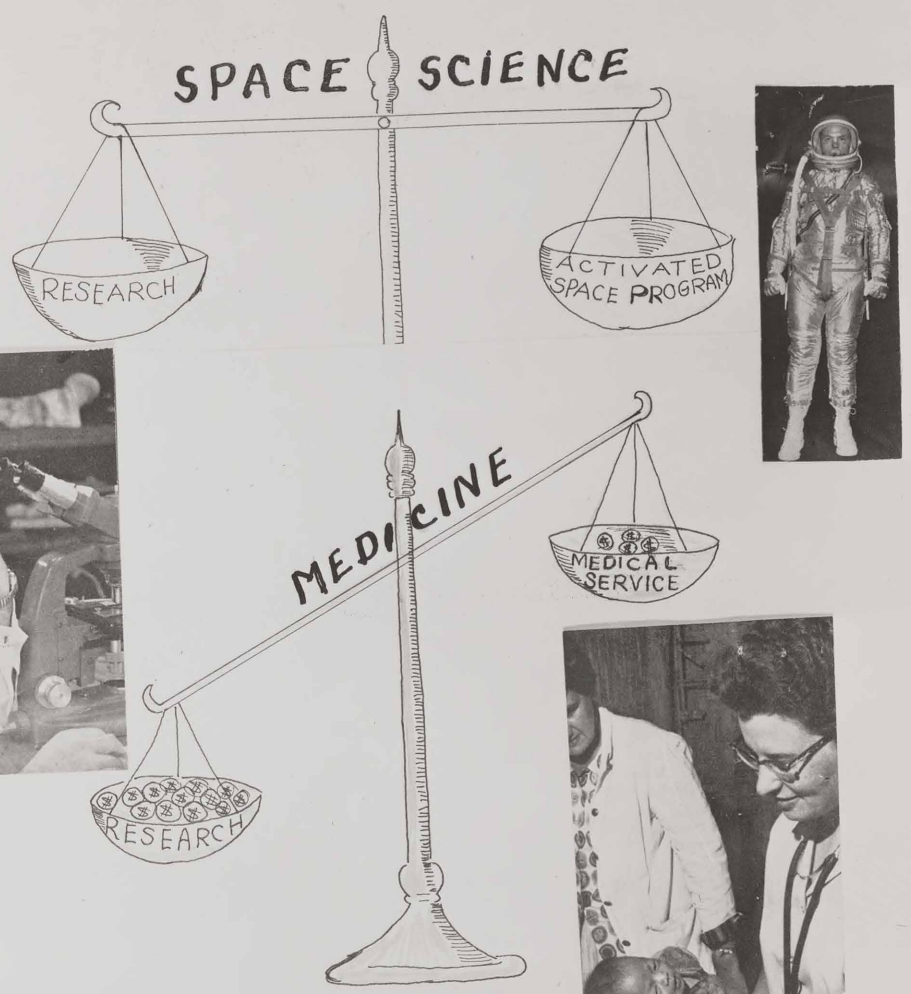


MOTOR DYSFUNCTION	HYPERKINETIC
HEARING DISORDER	EMOTIONALLY DISTURBED
LANGUAGE PROBLEM	VISUAL DISORDER
SPEECH PROBLEM	RETARDED
CONVULSIVE DISORDER	CONGENITAL ANOMALY

OUR CHECKER BOARD CHILDREN

24 picas

SLIDE IX



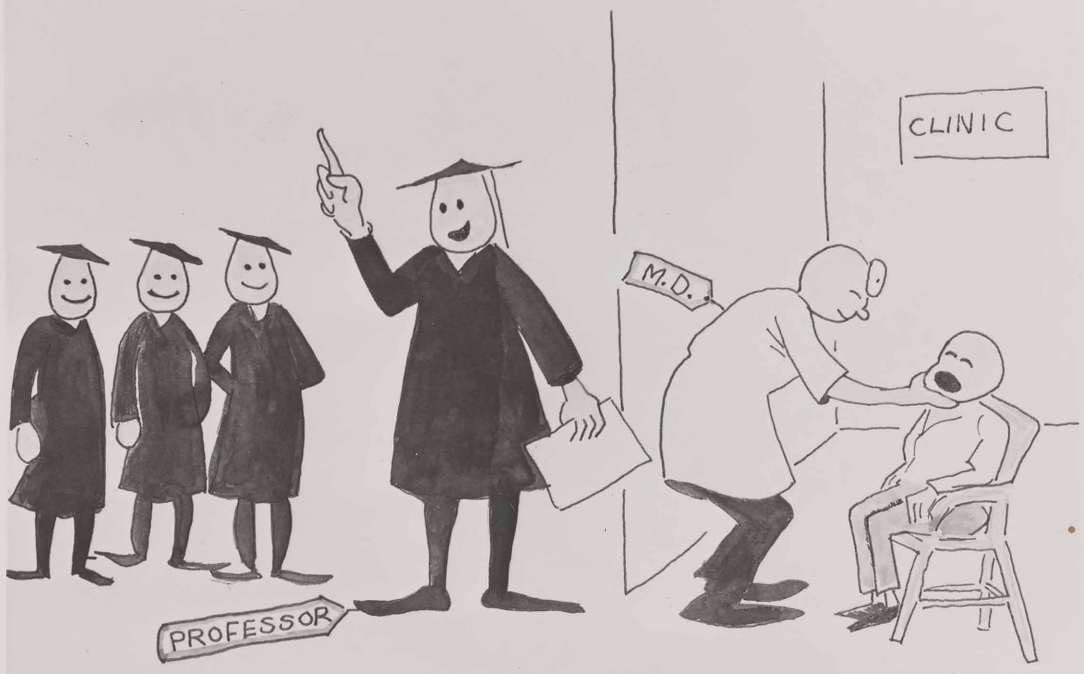
THE RESEARCH SCIENTIST AND THE CLINICIAN ALSO SHOULD BE COLLEAGUES

24 Picas

SLIDE X

EDUCATION

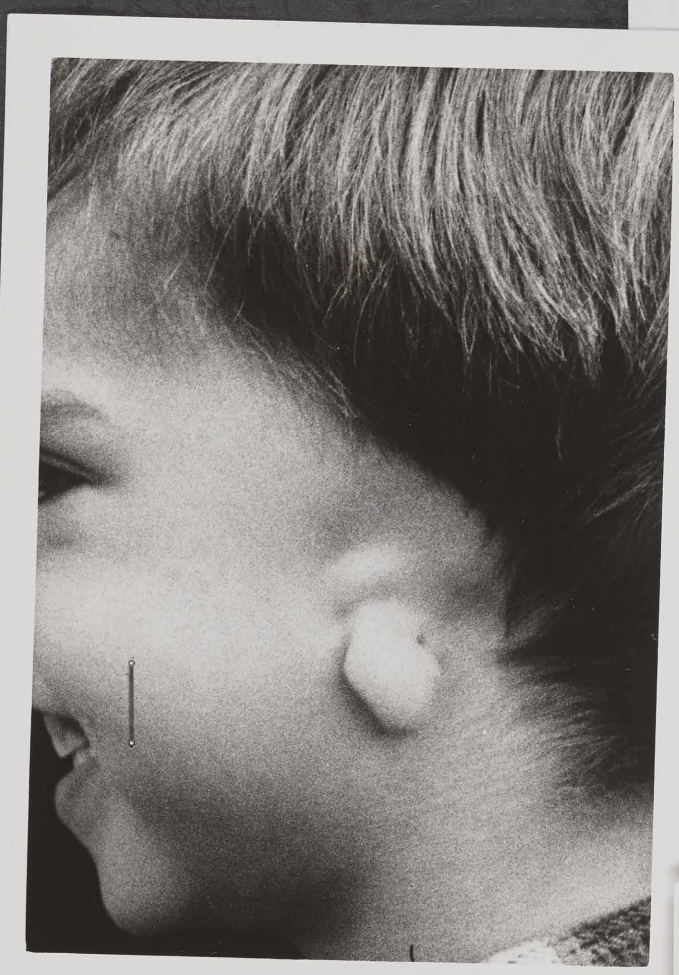
SERVICE



THE INSTRUCTOR AND CLINICIAN ALSO SHOULD BE COLLEAGUES

24 picas

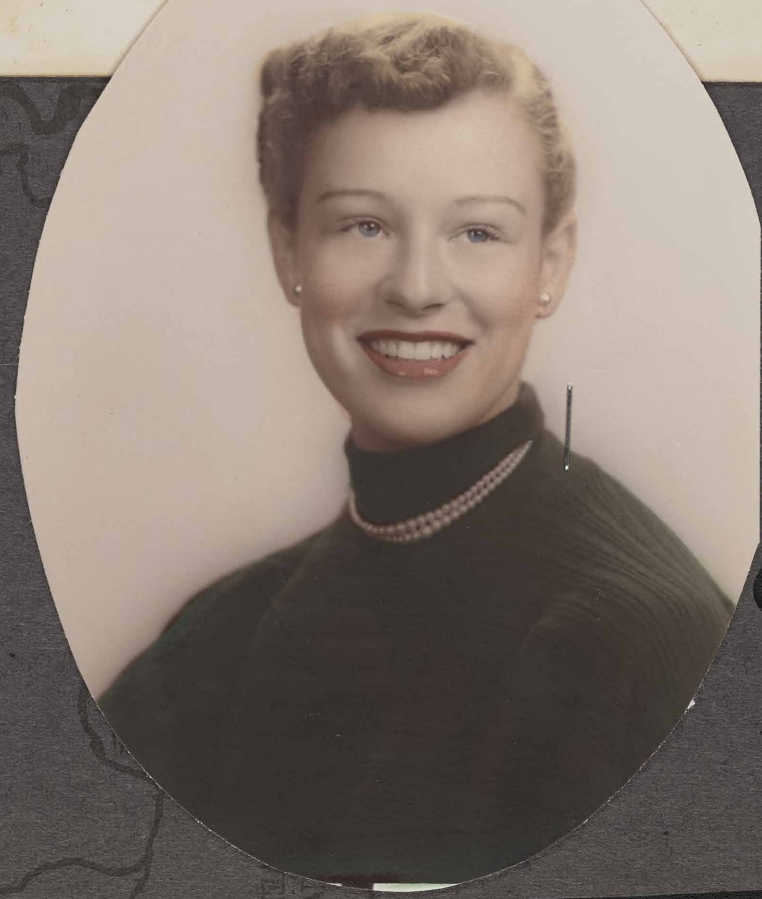
Marcella O'Neil



4/63 Ron Cole yes

T.V. Interview

To Dr. Ihelander
with all my love
"Pam"



Dear Dr. Joe.

'74 From Chris Cadden – his wheels !!

From the picnic 10/29/74

Cadden, Kehoe, Rosbuck, Grzadzo, Belotz
Newman, Jase Selge

Steve Kehoe in Hawaii 1974

"THANK YOU FOR ALL OF YOUR HELP"

"Dear Hospital Personnel:

Many thanks for all the help during the Auxiliary Race Day Drive, a Drive without you would be impossible.

At last count all of you sold close to 900 Books.

Again thank you for a successful year of close to $100,000.00—all of which goes for the benefit of Children's Hospital."

Sincerely,

Diane King

Diane King, President
Auxiliary of The Children's Hospital

INTER-COM OF CHILDREN'S HOSPITAL

VOLUME SEVEN
February

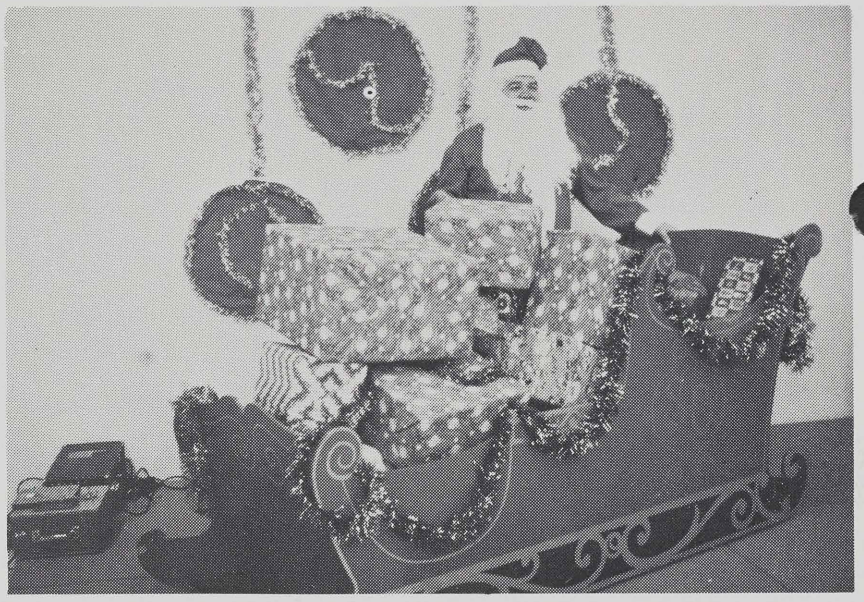
NUMBER ELEVEN
1974

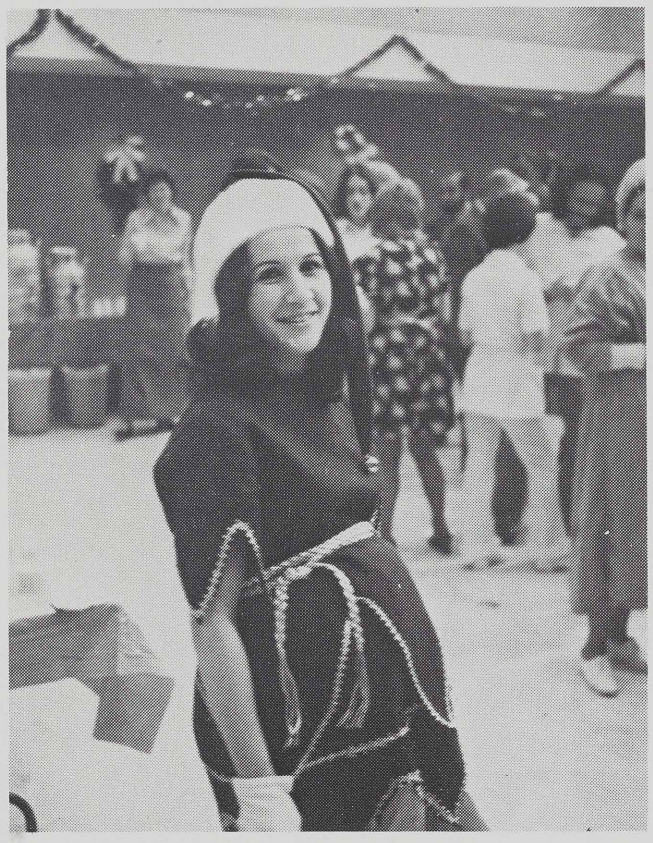
COMMITTEE: Jacqueline RR Carter-Walker, Editor; Ben Quarrels, Designer; Joseph Robertson, Photography; Barry Bastian, Special Features; Lucky Auzins, Production Control; Cathi Elias, Holiday Section; Eleanor Harrington, Clare Martin, Jane Musser, Rochelle Myers and John Wilson . . .

Robert C. Densmore, Administrator

CHILDREN'S HOSPITAL OF SAN FRANCISCO - 3700 CALIFORNIA STREET

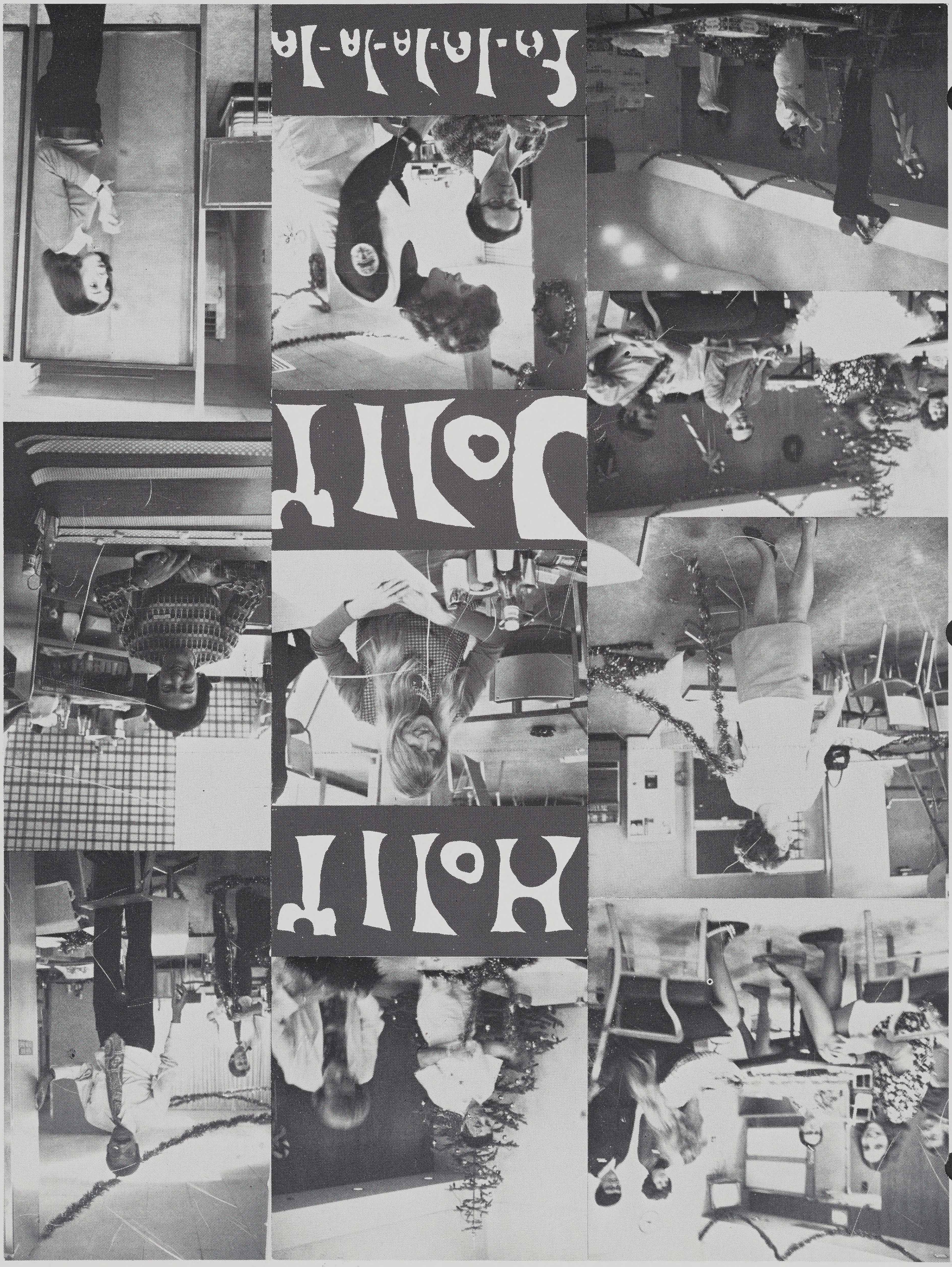
Christmas





is

for Children's


Fa-la-la-la-la
JOY
JOY

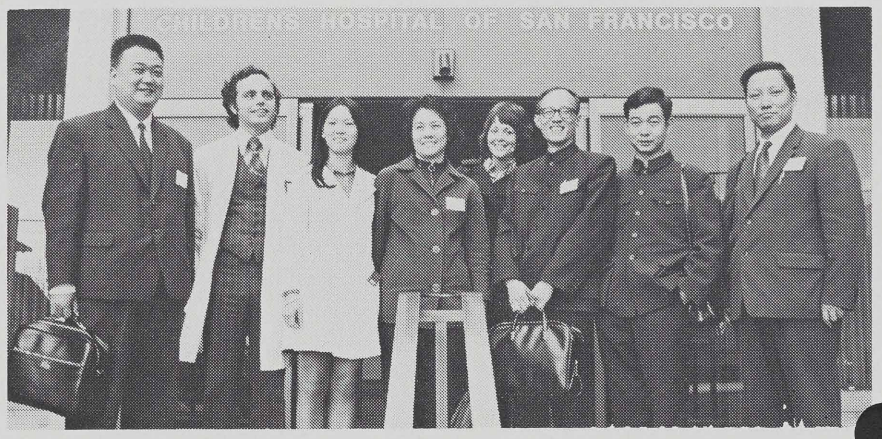
KENYA'S MRS. RUTH HABWE AND CHILDREN'S BOARD PRESIDENT, MRS. CHARLES LOWREY, VISIT ONE OF OUR PATIENTS.

One of Kenya's leading educators and advocates of women's rights, Mrs. Ruth Habwe, recently toured the Bay Area to meet outstanding local women active in professions, the arts, education and social service as well as to visit schools and hospitals, including Children's Hospital of San Francisco. She has been traveling throughout the United States under the sponsorship of the African Studies Association of Brandeis University and the Bureau of Educational and Cultrual Affairs of the U.S. Department of State.

Mrs. Habwe is the past president and one of the four founders of the Women's Progressive Society of Kenya. This organization, founded 28 years ago during the time of the British colonial rule, now has 50,000 members. For most of her life, Mrs. Habwe has dedicated herself to improving conditions for women and families in her country. She has traveled throughout Kenya, often flying with a bush pilot in a small plane to the more remote villages. Beginning with basic instructions in sanitation, nutrition and child care, she later expanded the program to combat illiteracy and encourage the creating of native artifacts. Her work with the women of Kenya's tribes was done in her free time and often financed out of her own pocket.

A head mistress, teacher, and developer of nursery schools, Mrs. Habwe was also a delegate to the 19th session of the United Nations General Assembly.

Mrs. Habwe's newest project is teaching African Creative arts and handicrafts in the villages for youth self-help projects under the Ministry of Commerce and Industry.

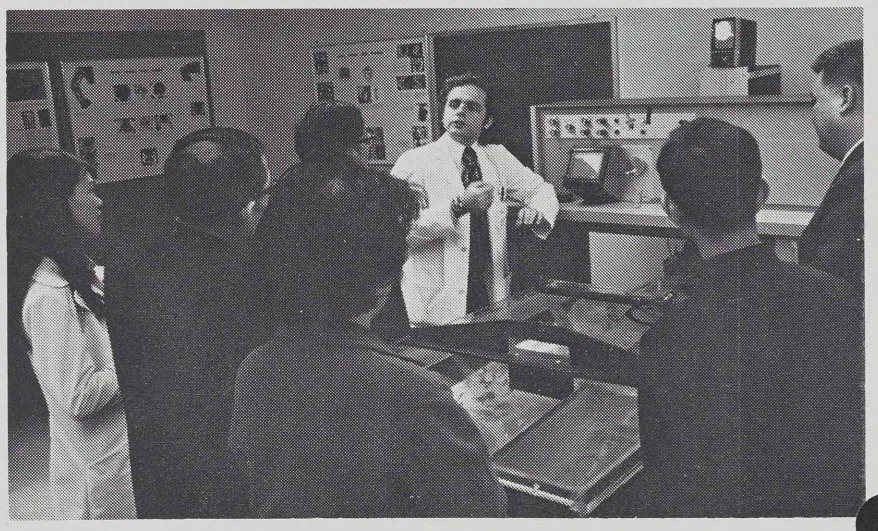


VISITING SCIENTISTS

Scientists from the People's Republic of China recently visited Children's Hospital of San Francisco. They were the third of four delegations that visited the United States as a result of exchange agreements reached in China.

The visits are sponsored by the Committee on Scholarly Communications with the People's Republic of China, the American Council of Learned Sciences, the Social Science Research Council and the National Academy of Sciences.

MRS. HABWE AND ASSOCIATE SHOWN IN DISCUSSION WITH MRS. HILARY FRUGOLI AND MRS. R. M. RANDALL.



MEMBERS OF THE CHINESE MEDICAL PROFESSIONAL GROUP WHO VISITED CHILDREN'S FOR A DISCUSSION OF ACOUSTICAL HOLOGRAPHY WITH DR. VICTOR RICHARDS AND DR. DAVID HOLBROOKE ARE SHOWN HERE IN A SESSION WITH DR. HOLBROOKE.

MOTHERS' MILK BANK RETURNS TO CHILDREN'S

The Mothers' Milk Bank, which has just returned to new quarters in the Sophie Pierce Brownell Laboratory in the basement of Children's Hospital, recently honored Mrs. Brownell one of the founders and an active participant in all of the bank's phases of growth and change, with a special tea and memories session of the history of the Milk Bank.

Now in its 25th year, the Mothers' Milk Bank was originally in the building which housed the Nursing School of Children's at 3801 Sacramento. When that was removed to build the Out Patient Research Building the Bank found new quarters at PMC. We are happy to welcome them back to Children's Hospital.

There were over 3000,000 babies born in California last year, the lowest birthrate since 1937. During that year, the Milk Bank collected 28,598 ounces of milk from donor mothers and helped over 200 babies. One of only two such banks in the United States, the other one is in Deleware, they have shipped milk as far as Hawaii, Texas, Arizona and Alaska.

MRS. SHEA, IN HAT, SPONSOR AND SISTER OF THE LATE MRS. ADOLPHUS GRAUPNER, ONE OF THE MILK BANK'S FOUNDERS AND MRS. BROWNELL SHOWN RECEIVING GREETINGS AND A HEARTY HANDSHAKE FROM DR. RICHARD LEONARDS AT THE RECENT TEA IN HONOR OF MRS. BROWNELL.

New officers to serve the Mothers' Milk Bank include: Mrs. Richard Leonards, President; Mrs. Verrel Weber, First Vice-President; Miss Olga Schwellinger, Second Vice-President; Mrs. Stephen Lazzareschi, Recording Secretary; and Ms. Joan Boothe, Treasurer.

All of the activities of the Milk Bank including recruitment and pick-up and delivery are carried on by the devoted volunteers with the assistance of two staff members, an office secretary and a nurse. New volunteers are always welcome.

* * *

Hirsch Handmaker, M.D., Co-Director, Nuclear Medicine Service, was the featured speaker for a series of nuclear medicine workshops and discussions sponsored by Wasau Hospitals, Wisconsin.

* * *

Dr. Handmaker was also guest speaker for the Mid-Winter North West Chapter Meeting of the Society of Nuclear Medicine. He spoke on the topic "Nuclear Medicine and Clinical Pediatrics," which is the title of a book to be published by Dr. Handmaker and Dr. Jerold Lowenstein.

* * *

* * *

David Holbrooke M.D. participated in the recent scientific session of the Eighteenth Annual Meeting of the American Institute of Ultrasound in Medicine at the University of Michigan Medical Center. He also presented a special film on Holography which was produced by the Medical Engineering Laboratory of our Surgery Department.

* * * *

WE GET LETTERS . . . .

"Dear Mr. Densmore:

Words fail to express our appreciation for all of the many kindnesses that your staff bestowed upon us during our stay in your hospital.

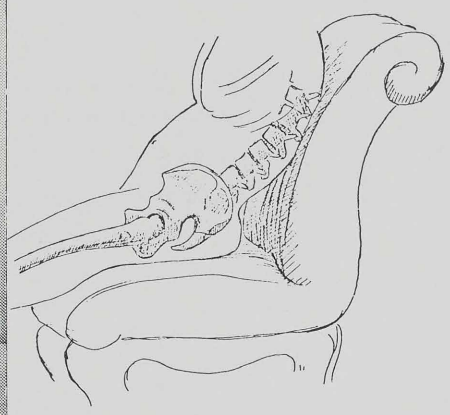
Strangers in a strange city and you took us in and did so much for us.

All three shifts on the 5th floor were just wonderful. They have excellent training.

May our Dear Lord bless each one of you for the wonderful work you are doing . . ."

HOW TO SIT TO SAVE YOUR BACK

Fourth in a series of articles by Mike Keropian, Chief Physical Therapist and National Director, Mesendieck System of Functional Movements . . .



How you sit affects the shape of the body and the health of the back.

The above pictures illustrate just one of the incorrect sitting postures which you see all of the time and which can cause a variety of back problems . . .

In the correct sitting position all of the bones are in proper alignment, each muscle is performing its proper function, and the body is in harmony. There is no undue tension, no undue pressure on any one part; no muscle is carrying more than its share of the load. Muscles and nerves are functioning smoothly, like the oarsmen in rhythm, smoothly gliding a shell.

Nine Steps in Assuming the Balanced Sitting Position:

1. Sit toward the front edge of the chair. Only by sitting toward the front of the chair can you assure proper use of the sitting bones. Otherwise you will be sitting partly on your thighs, drawing the back out of line.

2. Place both feet flat and parallel on the ground, a few inches apart. The feet serve as a good foundation for the body even in the sitting position. Have the feet flat on the floor, straight and parallel to each other and about three inches apart. The pressure should be directed at the big toe ball joint.

3. Use the sitting bones. (see illustrations) with some practice, you can readily know when you are seated on the balancing point of the sitting bones. It will be the spot where the pressure of the sitting bone is greatest against the chair and the pelvis erect.

4. Hold the knees parallel. With the legs bent, have the knees face straight forward and about three inches apart.

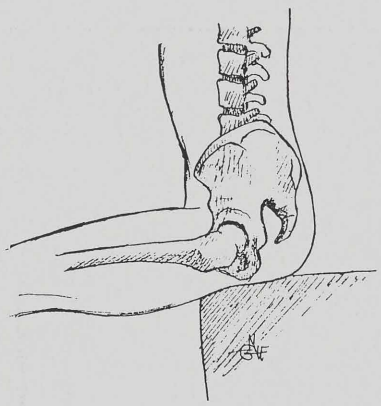
5. Press lightly onto the ball of each foot, and onto the sitting bones.

6. Draw the head and trunk up tall and straight. With the long back muscles, slowly stretch the spine straight upward toward the center of the head.

7. Hold the chin at right angles to the neck.

8. Gently, with the shoulder blade muscles, draw both shoulder blades toward the Middle Line.

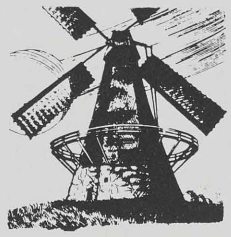
9. Let the arms hand loosely at the sides.



You have but to look in the mirror when you are in the Balanced Sitting Position to see the difference it makes in your appearance, you will soon feel the vast difference which proper sitting posture can make in your physical well-being. Start now to train your muscles for a healthier back and a more attractive body.

NEXT MONTH WE WILL FEATURE SOME ESPECIALLY BENEFICIAL EXERCISES FOR EMPLOYEES WHO SIT MOST OF EACH WORKING DAY . . . EXERCISES FOR THE HIPS . . .

Here Come The Windmills?



Is there a windmill in our future at Children's Hospital?

A Florida energy company foresees windmills dotting the U.S. landscape, with rotors 200 to 300 feet in diameter and extending 500 feet into the air.

The National Science Foundation is also interested in this approach, and will spend $1,250,000 in the next five years on its study.

A farmer of a 100-acre Des Moines farm has been using three big windmills to pump energy for his gamut of electrical appliances for 50 years. Batteries take up the wind's slack, and never a utility bill.

Another approach is that of a San Luis Obispo home that is heated by the sun via a sausage shaped water-filled bag which serpentines its way in 8-foot lengths back and forth across the roof. Cal Poly University is taking a serious look at this method.

WHAT CAN WE DO HERE?

CHIEF ENGINEER WILLIAM TERRY SUGGESTS THE FOLLOWING ENERGY - SAVING ACTIONS FOR EMPLOYEES:

1. TURN OFF LIGHTS NOT IN USE. TURN OFF FLUORESCENT LIGHT FIXTURES FOR LONG DURATIONS ONLY.

2. TURN OFF ALL APPLIANCES, INCLUDING ELECTRIC TYPEWRITERS, WHEN NOT IN USE.

3. KEEP WINDOWS CLOSED WHENEVER POSSIBLE.

4. KEEP THERMOSTATS AT 60°F IN UNOCCUPIED AREA (VACANT PATIENTS ROOMS) AND AT 68°F ELSEWHERE. KEEP RADIATOR VALVES OFF WHEN NOT NEEDED.

5. WALK THE STAIRS WHEN PRACTICAL, INSTEAD OF USING ELEVATORS.

6. REPORT LEAKING HOT AND COLD WATER FAUCETS AND RUNNING TOILETS.

7. REQUEST THE ENGINEERING DEPARTMENT TO REDUCE LIGHTING IN AREAS IF PRACTICAL.

Mr. Terry adds, "Of course most of this can also apply to ways to save energy in your own home . . ."

MORE ENERGY SAVING IDEAS NEXT MONTH

CARBON MONOXIDE POISONING— A DANGER OF THE ENERGY CRISIS

The current energy crisis may be accompanied by an increase in carbon monoxide poisoning. Fuel shortages will spawn the use of various devices for providing additional heat in homes. Fireplaces and other heating facilities which have not been used recently may be placed into operation. Catalytic heaters, charcoal burning units, and similar devices may be used indoors or in confined spaces. Many individuals will undoubtedly resort to "makeshift" measures to prevent the loss of heat from their homes, including installing additional insulation, sealing cracks and crevices, covering windows with plastic and using other measures which tend to make the home "airtight". Any measure that reduces the amount of air movement into a home, coupled with a defective heating device or the improper use of a combustible fuel indoors enhances the potential for poisoning by carbon monoxide. (excerpt, HEW)

"All mankind is divided into three classes, those who are immovable, those who are movable, and those who move".

B. Franklin

THE BOARD OF DIRECTORS, ADMINISTRATION, MEDICAL STAFF AND THE ENTIRE FAMILY OF CHILDREN'S HOSPITAL OF SAN FRANCISCO WISH TO EXPRESS DEEP SYMPATHY IN THE DEATH OF MR. SILER B. (SI) STEELE . . .

Mr. Si Steele

Purchasing Agent For Children's Hospital

1959 - 1974

12/18/1964

The Child Development Center was opened August 1, 1952 under a grant from the United Cerebral Palsy Association of San Francisco. The Child Development Center now sees any child with a handicap referred to it by a physician or medical facility. This has been made possible by support from the following sources:

1. United Cerebral Palsy Association
2. CrippledChildren Services
3. March of Dimes
4. Special donations

Recently a fee for services has been instituted.

In order to assign the patient to the proper category for financing stamps are used such as C.P., CCS, Congenital Anomalies, etc.

Cerebral Palsy requests a yearly report. The March of Dimes has requested a report every six months. Crippled Children Services expects a report after the study of the child has been completed and the conference held.

During the year 1963 268 new patients were processed.

In 1963 the Cerebral Palsy Nursery School had 36 children enrolled in the morning and 53 children were seen in the afternoon. There were 28 psychological tests on these patients. Thirty three children had speech therapy. One hundred forty one had physiotherapy and seventy eight had occupational therapy. One hundred twenty nine were seen in the medical clinic with 23 new patients.

In the Birth Defects Center for the same period there were a total of 245 new patients and a total of 464 patients attended medical clinics. The children in the Birth Defects Center came from twenty different counties and some from the State of Nevada. A very high percentage in both groups have either some degree of mental retardation or a major learning problem that limits performance. The great majority of the children have had complete evaluations including psychometric tests. The extent of evaluation depends somewhat on the age. They all have developmental studies and most of them, unless they are infants, have an educational evaluation. Many have speech and hearing evaluations. Psychotherapy and occupational therapy also see a great many of them, at least for evaluation. Inasmuch as many of the Birth Defect children come from long distances the therapy for these must be arranged in the local community. This involves the social service department to a very great extent and last year our social worker saw 233 families. The past few months we have had an additional social worker to help us.

This type of service represents an almost overwhelming amount of work, there is a tremendous demand for reports which go to referring doctors or agencies, often to school facilities, sometimes to a special treatment center such as the hearing or speech center, often to a psychiatric or mental hygiene clinic. A fair number of the patients are referred for State care at Sonoma and a few to Napa.

The policy of the Center has been to serve the patients and their parents. This is our foremost concern. With the increased interest at both the State and National level in children with mental deficiency and deprivation there has been a great encroachment on our time. We actually have a tremendous number of visitors, they literally come from all over the world, apparently we are on everybody's list - local, State and National. Educators in many areas bring students over; nursing schools similarly want to bring the nurses over to visit. Members of the staff of the Center have often been called upon to participate in community, local, State and Federal programs for the handicapped. Long detailed reports have gone to the President's Commission on Mental Retardation, to the Governor's Commission

and to several other study groups. The final reports of these Commissions which are now just beginning to come in are so much in line with our own analysis of the problems that we feel our efforts have not been in vain. Members of the staff have also been called upon to speak at various groups at local, State and National levels.

Much information has accumulated that should be documented, analyzed and published. The work has also resulted in a great many questions that should be further investigated. At present Dr. Helen Pryor is volunteering her services for a study of growth patterns by using anthropometric measurements. She has detailed curves and means on 6,000 normal boys and 6,000 normal girls. We have already done the measurements on over 100 mongols and about a dozen on Turner's syndrome. We also have measurements on many of the children with congenital defects or handicaps from hypoxia or trauma. A second project contempated is a study of twins. Dr. Gilbert Webb has outlined a study of twinning at Children's Hospital over a period of 10 years; this means that we have over 300 sets of twins that we wish to study. A preliminary review of this project has been made. In addition to this we have some 40 or 50 twins in our program where either one or both are handicapped. Recently I reviewed 100 random cases for an analysis of the prenatal factors that might have contributed to the pathology. It was interesting to find that 16% had either definite hereditary anomalies or the family had many anomalies in the parental histories. Some 34% were attributed to maternal factors involving health problems and another 30% to intra-uterine factors. A relatively small percent were due to birth injuries.

LOOKING TO THE FUTURE

There is a segment of the population that is less than perfect. They are born, they grow up and they create problems. This is a medical problem primarily as these children continue to have long term chronic handicapping. It is up to the physicians to attack it in much the same manner as they have attacked other major hazards both in treatment and prevention. We will not find a vaccine or immunization procedure to prevent these cases from happening but a great deal can be done to tighten up our defenses against the injury to the foetus. This pertains particularly to the use of drugs during pregnancy, avoiding infection during this period, radiation and so on.

The physicians to date have volunteered their services to the program. The director now gives over one-half of her time. Sometime in the future it will be necessary to have a paid director. There are many unanticipated costs that arise in the program. Some of these are:

1. Transportation - A taxi book costs $5.00. This City has never gotten around to provide any transportation for the handicapped, except for public school programs.
2. Patient Equipment - Wheelchairs, crutches, and other home equipment often must be purchased as an emergency. The official processing often is slow and tedious. Any sum from fifty to several hundred would be a help in this area.
3. Similarly laboratory procedures are often desirous although not absolutely necessary and these are expensive for the family. It becomes almost impossible to process these needs through any official agency.

This applies particularly to such studies as chromosome counts or electrophoretic patterns in an entire family. Several hundred dollars could be used for this, but even any small sum would help.

4. Our research is done entirely on a volunteer basis. A grant has been requested - the first one submitted was turned down. This happens frequently with grants. Sums from one hundred to several thousand dollars could be utilized.

5. A much needed fund is one to hire someone to put all our information on IBM cards so we could analyze our findings. The coding should be done by a physician. It is estimated that $3,000 or $4,000 would be required.

12/18/64

ORGANIZATION OF A NURSERY SCHOOL FOR THE PHYSICALLY HANDICAPPED

A GUIDE

A Nursery School for handicapped children has been conducted at Children's Hospital for over twelve years.

The following guide for establishing such a nursery school is based on this experience.

SPACE

A fairly large area for a playroom is desirable; our present room is 17-1/2 feet by 20 feet.* Young children with physical defects will need physical and occupational therapy; there should be separate rooms for these activities, not less than 15 feet by 10 feet. Separate rooms are desirable since these children are often distractible and it is essential that noise and confusion be kept to a minimum. Equipment for the therapy room should be selected by the therapists themselves, and there should be storage or cupboard space within the treatment rooms to reduce the clutter that is distractible to the children. One can, however, improvise by screening or curtaining off treatment areas from the main playroom. In assisting other groups to start we have found it helpful to them to point out that it is important to avoid having parents and children enter the playroom directly; if there is not a hall adjoining the playroom then an entryway can be improvised with screens so that when the children do enter the playroom they are ready for the morning's activities. Lockers or coat racks for their clothing can be in the entry way to avoid confusion at entry time and also at the end of the session.

Additional separate rooms will be needed if the program includes other professional staff: speech and hearing consultant, psychologist and social worker. An office for secretarial help and a small office for the nursery school staff is necessary. We feel that it is essential for a social worker to be identified with the program from its inception. Her office should be accessible to the playroom but should offer some privacy for the parents being interviewed.

If at all possible, though expensive, a viewing room with one-way windows adjoining the playroom is helpful; such a room should be large enough to accommodate the mothers who are meeting with the teachers or the social worker during the nursery school session. Access to the viewing area should be controlled so that the staff is aware at all times when the room is in use, by whom and for what purpose. Since most beginning budgets do not permit such an item as a viewing room, perhaps an area of viewing glass could be placed in the playroom door; in this way the confusion and disruption of visitors in the playroom can be avoided.

Storage space is usually at a premium and one may need to use space not accessible to the main playroom. Many programs are required to store all equipment at the end of the session. Wheelchairs, walkers, relaxation chairs stored in corridors often pose a safety hazard.

* At present we have 27 children enrolled, some attend three and others two days per week; ten attend each morning; our present space is <u>not</u> adequate.

Isolation areas for children who become ill during the morning can be made with screens. Toilet facilities are often a problem. Adult size toilets can be adapted by making wooden steps leading up to the toilet, wooden hand rails can be made for the lavatory and wooden planks which are removable can be used to convert steps into ramps if needed. Shelves in the lavatory can be made to hold wet and soiled clothing. A safe, sheltered and fenced-in outdoor play area is most desirable, and is often a local requirement for licensing, but if not available children can be taken for walks and excursions during the school sessions.

STAFFING

Many programs begin with a nursery school teacher (who should be trained in this field) and volunteer help. The teacher should be responsible for orientation of these volunteers on nursery school techniques and procedures and she should be cautious about over-preparing her helpers so they do not become unduly anxious. Volunteers and mothers working directly with the children should do so only with preparation. Such preparation should include talks from professional people in the community, medical people for example, who will discuss various handicapping conditions and their implications. This is an important aspect of developing a good program.

It is our feeling that the initial staff could consist of the nursery school teacher, the physical and occupational therapist. If the funds are limited and the initial enrollment is small, staff can be used on a part time basis. In even a small activity, the psychologist, the speech-audiologist and the social worker should be available for consultation. Our medical staff, including our medical director who is a pediatrician, as well as our orthopedic and neurological consultants, give their time on a volunteer basis.

Staff as well as volunteers working with the children should have a "pre-employment" physical examination which should include a chest x-ray and these should be repeated yearly. The regulations of the State Department of Social Welfare for licensing nursery schools are an excellent guide for the matters we are discussing.

Any physical plant, however minimum, CAN provide an effective program when oriented toward patient service and when supported by interested parents and a community whose goal is to provide services for children with special problems.

A nursery school for handicapped children needs medical supervision, preferably by a pediatrician. An adequate amount of medical information, including at least a diagnosis, is essential information for the staff to have. Physical and occupational therapists always do their treatment as prescribed by a physician.

A social worker can function as liaison between medical staff and nursery school staff as well as between medical staff and parents; this is in addition to her counseling of parents directly. Psychiatric consultation is invaluable in any program working with parents of the handicapped. Such consultation is no less essential to the staff working with handicapped children and their families.

A nursery school can be a place of socialization for the children as well as a place for therapies; it also offers continuing observation which leads to a better assessment of a child's functional abilities. The staff is then in a better position to offer parents counseling in planning for the child's next step whether this be another school, foster home care, or institutionalization. The parents' real emotional acceptance of the child's limitations as well as his assets is often a long, slow process. We need to help parents cope with tomorrow before we can help them begin to plan for next year.

CRITERIA FOR SELECTING THE CHILDREN

Beginning groups are confronted with many decisions; how many children can be provided for? What kinds of handicapping conditions will be accepted? What should the age range be? We have found that defining any policy too rigidly is not advisable and may exclude the very child the program could help. However, often nonprofessional advisory board members will need interpretation by the staff as to real limitations of their services. For example, a severely physically handicapped child who is not ambulatory should not be in the same group with an older, active child whose main problem might be his mental retardation. The age span should be flexible in both upper and lower limits, for in stature as well as in developmental level some of these children are below their chronological age. The amount of special attention children often require from the staff is a determining factor. We have recently divided our children into two main groups: three mornings a week we have the physically handicapped who have potential for going into another school program; two remaining mornings a week we have the children who are younger, who function at a lower intellectual rate, or who are new to the group and we have not yet determined their functional level, or feel that their diagnosis is not yet determined. The groupings are flexible and the children are moved to different days whenever indicated. We feel that some less able children do better when placed in a group where there is less competition, less activity, and less confusion. The nursery school can provide a respite from the ceaseless competition some handicapped children face in their environment. As a child grows and changes, his needs also change and we feel free to try him in another group.

Organizing a nursery school will point out the need in most communities for still another service: that is, a day-care program for the severely mentally and physically handicapped child. Such programs as these could be staffed by matrons, under professional guidance, and can offer physical and perhaps occupational therapy. These programs should be geared to offering relief for the mother and a change in environment for the child, rather than unrealistically seeing it as preparation for the next school step.

It has been our experience that many parents regard acceptance into a "nursery school" as implying trainability or educability, and this may not always be the case. Indicating in the beginning, in the intake interview, that entry into the group is on a visiting or trial basis, saves the parents' feelings of disappointment and failure if it is found, after careful observation, that the child will not profit at this time by attending the group. It is essential that the child's entry into as well as his termination from the group be made jointly by the medical director and the nursery school staff. The physical safety of the handicapped

child and the amount of physical care he requires indicates that the groups should be small enough so that each child is well cared for. He may need picking up, carrying, changing and feeding and the staff should be large enough to attend to each child's needs. We have felt that ten children each morning with a staff of four adults insures individual attention to each child. Handicapped children are, in reality, dependent, and the nursery school may be their first experience outside the immediate family circle, often their first exposure to other children, and the staff should respond to their needs in a warm and reassuring way, especially during this initial adjustment period.

New children should attend about an hour their first days with the length of time gradually increased as they are able to tolerate it without fatigue. We require mothers to remain with the children until we feel the child is ready for the separation (often the child is ready before the mother) and this we do not rush. These children are dependent and growth toward independence needs to be at their rate, not ours. Parents who want to "just slip out" of the playroom without telling the child are not permitted to do so. Consultation with the social worker on the meaning of separation for the child has proved invaluable to the parents and is then reflected in an increased ability to "let go". It is helpful to point out to new parents that the children should come to nursery school dressed appropriately so that they are comfortable about crawling, playing with water, finger painting, etc. We have recognized the needs of some parents to have their child "dressed up" for school; in such cases we merely suggest that comfortable clothing can be left at school and we will change the child so that he can enjoy the mornings activities.

EQUIPMENT

If it is needed, have professional help in selecting chairs and tables that are the correct height for young children. Be sure that all surfaces are washable; large washable mats for floor activities or for therapies are important; if space is a problem, mats can be hung from the wall when not in use. If the budget does not permit lockers for the children's clothing, safe blunt hooks nailed to a board and well spaced can be nailed to a wall surface. Be sure they are the right height for the children. Again we stress having such hooks or lockers in the hall or entry if possible.

If the space is small and funds are limited a piano for the playroom may be a luxury. Record player, thythm instruments, auto harp can be used instead. If space does not permit a jungle-gym for climbing (this may be somewhat limited in use with handicapped children) a small, safe slide which can be dismantled for storage can be made by parents. A doll corner, a dress-up corner can also be made and equipped by parents; have the occupational therapist help design simple doll clothes that are easy for the children to put on the doll. Avoid donations of doll clothes that are difficult even for adults to put on. Blocks are expensive, but an initial investment of safe durable blocks may be something a generous donor would consider. Cardboard building blocks have been successful with handicapped children since they are light in weight. Smaller matched sets of wooden blocks to be used alone or in conjunction with small cars and trains are useful. Fathers can be given specifications for a durable, doublesided wooden easle with a tray

Nursery School for Handicapped

designed to hold the paint cups firmly. A sturdy doll buggy can be used in play and in therapy by adding sand bags of appropriate weights. Cabinets with doors that lock are useful for storage of paints, and other supplies in the playroom; boxes made to fit under the cabinets give extra storage space for toys. Plastic lined smocks for the children to wear while finger painting or for water play can be made by the mothers; again, keep them simple and easy to put on and take off. Terrycloth bibs with plastic lining are items used a great deal and can be made by mothers. Fathers can make racks to hold the puzzles, which should be simple and have durable washable surfaces. Playschool and Creative Playthings catalogues offer ideas. A set of farm animals and jungle animals made of rubber and bought commercially are an excellent investment.

Economy should not be practiced when buying the art materials for the children. Good brushes should be selected with the occupational therapist's help in determining varying length of handles; large crayons, poster paints and finger paint in small jars so the children can help themselves are essential. Books should be carefully selected; better a few good books than many cast-offs, "donated" books. Phonograph records should be carefully selected, and again it is well to be temperate in accepting donations of records that have been outgrown by older children. Be sure toy cars, trains, etc., do not have sharp edges. Because many children will put many things in their mouths, be vigilant in removing small swallowable items from such things as Tinker Toys, etc. Size, safety, durability and washability is a good guide for purchasing equipment for a nursery school.

Special therapy equipment should be selected by the occupational and physical therapists. Therapy rooms should be free from the distractions of too much equipment. One of the most used items of equipment in our nursery school is a low, stainless steel sink (height: 22 inches) with cutouts which permit children in special chairs to reach the water. Sponges, egg beaters, are favorite water play toys.

Each community will have slightly different needs to meet. In the beginning there may be only three or four children, and a church basement may provide space. It is important to begin with a general plan that is not too rigid. Through experience gained in working with handicapped children one develops criteria that are realistic in terms of space, staffing, budget and other community services. The important thing is to <u>begin</u>.

Jane Sledge, Director, Nursery School Program
Child Development Center
Children's Hospital, San Francisco

COMMUNITY ORGANIZATION TO HELP THE HANDICAPPED - A GUIDE

There is increased public awareness and concern for the poor, the deprived, the unwanted, the retarded and the physically handicapped. This, coupled with the allotment of funds from State and Federal sources for Aid to Communities, calls for coordinated planning at local levels.

Experience gained in the Child Development Center at Children's Hospital in San Francisco is presented here as a guide for those who may be initiating such service.

The focus must always be on the child, his family and their needs. The focus must not be deflected to organizations, agencies or staff personnel.

Local experienced people whenever possible should be enlisted in the planning. All existing facilities and agencies in the community should be fully utilized, adapted and expanded rather than creating new organizations. New organizations call for additional superstructure and increased expenditure which leeches the funds without bringing necessary aid to those who need it.

The basic needs of deprived, retarded or handicapped children are the basic needs of all children, namely a home and the love and security that goes with it, medical care, education, training for employment in adult life, recreation and the right to worship.

HOMES: In infancy and early childhood there is no perfect substitute for the child's own home and his own parents. This is the age during which the child learns trust, love, truth, right and wrong, the role of parents and siblings. If there is no home or parents the next best substitute is a good foster home.

After the age of 7 or 8 years the child becomes more involved with his peers. At that age, therefore, separation is better tolerated. If a child has a severe handicap such as deafness, blindness, cerebral palsy, or spina-bifida, or a combination of these a special class or a boarding school for 5 days a week may be a necessity. The weekends, holidays and vacations should be spent at home.

The child who is severely handicapped often both physically and mentally may need a semi-hospital situation during much of his childhood. He may need nursing care as well as rehabilitation services and this often cannot be adequately provided in a home situation with ambulatory care.

After the school years the adult with a severe handicap will need living arrangements especially adapted for his needs such as entrance without stairs, elevators with doors wide enough to accomodate a wheelchair, ramps, personnel in the building trained to assist and transportation.

MEDICAL CARE: Medical care is the service now most adequately provided to handicapped children. In addition to private care, there are hospital and university medical clinics, rehabilitation services and Crippled Chilren Services available for a wide category of diseases. Unfortunately total evaluation of the child not only medically but also psychologically and educationally is not always available in medical centers. A Child Development Center in which such comprehensive evaluation is possible should be available to all practicing physicians.

EDUCATION AND TRAINING: The public school provide many services for handicapped children. There are now classes for both the trainable and educable children, there are special provisions for the deaf, blind, the neurologically handicapped, the emotionally disturbed and the physically handicapped. Even with this impressive array of services there are large gaps. The public school does not provide facilities for the pre-nursery and nursery school child who is deaf, blind, retarded, aphasic or who has multiple handicaps. Recent studies indicate that 50% of intellectual growth takes place

by 4 years of age and 80% by 8 years of age. Provision for this education should be assumed by the voluntary agencies until such a time as public funds are available.

There are some specialized State schools but these also tend to take only the so-called "normal" deaf, blind or aphasic children.

The larger communities may have sheltered workshops for older handicapped children and adults.

Experimentation with work areas in small industries should be explored.

Employment of the handicapped needs to be kept under constant surveillance. In Sydney, Australia the Spastic Society purchased a plant that manufactured electronic equipment and the entire factory is now run by spastic individuals.

RECREATION: Recreation for the handicapped has been almost entirely neglected. This may be the easiest activity to initiate. Recreation can serve children who have a variety of problems. Various agencies can be pursuaded to participate in the program both with funds and with volunteers.

There is available an excellent book which describes in detail the methods for instituting a recreation program, namely "Recreation for the Physically Handicapped" by Janet Pomeroy, Macmillin & Co.

WORSHIP: The churches and synogogues should be sensitive to many needs of handicapped children and their families. There are some church-supported homes for the retarded but there should be many more. Very few churches have adapted their religious instruction to suit the handicapped whether he be deaf, blind or retarded.

The church building is often vacant most of the week and the space for some of the above described activities could be granted.

METHOD OF PROCEDURE: It is necessary that someone take the initiative in organizing a community for action. In the case of the handicapped child the most appropriate and logical person to assume leadership is the physician. Such a child is usually under close medical supervision all of his life - no other discipline or individual has this type of longterm contact with the child. The move therefore to organize the community might start in the County Medical Society or in the Pediatric Society, it might be instigated by the Public Health Department.

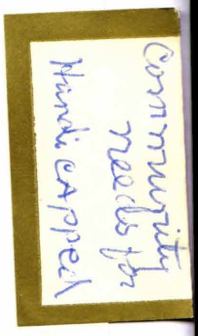
The existing community services should be listed and participation invited. Representatives from the medical community should meet with local leaders in public health, welfare, education, religion, recreation and so forth several times for open discussion and clarification of roles. Outside experts in various fields might be invited to participate such as from Public Health Services, Crippled Children Services, Centers concerned with child development, educational centers and so forth.

After a few general sessions for organization and clarification smaller committees should be formed to pursue special needs. If discussion continues without action for a prolonged period there may never be any action. This is particularly true if the group first resorts to applying for Grants and waiting for financial aid. Grants can be a delusion. If a Grant is obtained the usual procedure is first to hire a director and secretarial help and rent office space, a second step is to make an elaborate survey and then the third step is to re-apply for the Grant because the original one is nearly exhausted. It is better to start an active vigorous program that is woven into the existing fabric of the community. Local sources can be tapped for financial aid and when the needs are clear

outside funds can be sought.

The logical subdivisions for action are:

1. Nursery schools and pre-nursery schools
2. Day Care homes
3. Counseling
4. Recreation
5. Church activity
6. Parents groups
7. Legal rights and benefits
8. Transportation
9. Employment; including vocational testing, guidance, training and placement
10. Comprehensive diagnostic and rehabilitation services



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THE CHILD BORN LESS THAN PERFECT

Information for Parents

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THE CHILD BORN LESS THAN PERFECT

Information for Parents

Babies are living beings. They are conceived; they develop in the uterus (womb); they are delivered. They grow into childhood; then into youth. They mature; they reproduce; they age and die. All living things go through somewhat similar cycles. A grain of corn is planted in the soil, it sprouts, grows, bears fruit, ripens, ages and dies.

The farmer, the fruit grower, the gardener selects good seed, plants it in good soil, hopes for a favorable climate and expects a good yield. Sometimes things go amiss. The dairyman, the sheepherder, the dog fancier selects and rears his animals with a somewhat similar purpose. Sometimes a runt appears in the litter. In plants, animals and humans a certain number of the young are born defective.

When a child is born with a defect parents, relatives and physicians all ask "why". If we follow through the birth process again as if a film were re-run in slow motion SOME of the answers will be found. Some defects are inherited; that is, there is a fault in the seed itself. Most of us have heard about such defects through our history books. We may recall the stories of hereditary diseases in royal families such as hemophilia, the bleeding defect. This defect was found in many royal heirs, notably in Spain and Russia. There are other well known inheritable conditions, phenylketonuria, is one. There are several others. Most of these are rare and usually parents do not hear about them until a case occurs in his own family, then suddenly friends, relatives and neighbors all cite instances. There are other defects, the inheritance of which is less well understood. In some families there have occurred at intervals but fairly regularly such defects as club feet, harelip, cleft palate, missing or extra fingers, webbed toes, and numerous other abnormalities. Some inherited traits do not appear unless two people with the defective trait marry. These diseases are said to be "recessive", or hidden. Other conditions are inherited as "dominant", then, according to Mendel's law, three out of four children may show the trait. It is the above tendencies in inheritance that have led medical people to advocate that cousins or near relatives not intermarry. If they do these traits are more apt to be present in both parents and appear in the children.

In some conditions the entire pattern of the child is unlike the parents. This is the case in mongolism. Mongoloid children look more like each other than they look like the other members of the family. In 1954, Lejeune in France was able to grow human tissue and study the chromosomes, the hereditary particles of the cells of the body. He found that mongoloid children have 47 chromosomes instead of the usual 46. Chromosomes may be looked upon as tiny building blocks. Normally 23 come from the father and 23 from the mother, thus making the 46 in all. In the case of mongolism, one of the pair splits, or "twins", making 47 chromosomes in each cell. The pattern of growth, then, is different. It is as if you were knitting a complicated pattern and picked up an extra stitch. Your final design would be different from the original pattern. The mongoloid child is different. Since all mongoloids have 47 chromosomes, they resemble each other more than they do the other members of the family. An effort has been made in recent years to drop the word mongolism and instead to call it Downs' disease or syndrome after Dr. Downs who first described it. Another term used for this condition is "trisomy". This latter term refers to the three instead of two chromosomes in one of the 23 pairs normally found in humans. If the words, Mongolism, Downs Syndrome or Trisomy are used they all mean the same thing.

In another condition known as Turner's disease there are only 45 chromosomes or building blocks in each cell. Again, people with this defect look alike. When you are aware of this condition it is easily recognized. Since the discovery of chromosomes and other new techniques for the study of individual body cells, many variations have been

found. In almost every instance where this occurs the body form or design differs from the usual human design. In some cases a defect can be detected in the chromosome of one or both parents. This may favor the occurrence of defects in children born to them. Such a parent is said to be a "carrier" of the trait.

Mongoloids are mentally retarded but among their own kind they vary. The degree of retardation cannot be estimated by the number of defects the mongoloid has. For example, one mongol may have a congenital heart disease, crossing of the eyes, and flat feet but be brighter than a mongol that has none of these three defects. A good looking mongoloid may be more retarded than one with many physical defects.

In certain conditions the child worsens unless the defect is detected early. This is true in phenylketonuria or PKU and it is true in cretinism or thyroid deficiency. In the case of phenylketonuria the child is placed on a highly specialized diet low in phenylanalin, one of the essential amino acids in our food. On this diet he can develop more nearly normally than without the diet. The child gradually gets worse as the phenylaniline accumulates in his body and affects the normal growth of the brain. In hypothyroid infants or cretins the missing product from the thyroid gland must be supplied to the infant. If it is not supplied the brain fails to develop normally and the child becomes increasingly retarded.

It is an observed fact that a child born with one defect often has others. Even with the earliest diagnosis and treatment a fair number of children born with severe and basic defects such as cretinism or phenylketonuria do not attain the highest intelligence. In an organism as highly developed, intricate and complicated as the human being it is remarkable that defects do not occur more often than they do. It is also not surprising that many different types of defects may occur. There is no part of the body that may not be abnormal at birth. This is true not only for all the large and visible parts of the body but it is equally true of the internal organs, the heart, the kidneys, the intestines. It is also true for the chemistry of the body and the chromosomes and very likely for many still undiscovered mysteries of human development.

In recent years two events have focused attention on the developing embryo or fetus. Dr. Gregg in Australia noted that after a German measles epidemic many babies were born deformed. This observation has been confirmed around the world. Scientists therefore are trying to prepare a vaccine to immunize against German measles, hoping thereby to eliminate the possibility of a mother having the disease when she is pregnant.

In Europe a few years ago there suddenly appeared a great many infants born without arms or legs. After much doubt and debate a new drug called Thalidomide was implicated as the cause of these deformities. These two tragedies, German measles and Thalidomide defects aroused physicians' concern about the embryo and the fetus. Prior to this, doctors had tended to disregard drugs and diseases as having any influence on the fetus. Our disregard for such events probably stemmed from the fact that "old wives tales" regarding "markings" of the infants had been carried so far afield that it was discarded as of no creditibility. As happens often the pendulum tends to swing too far in the opposite direction. It probably has been taught too widely that the fetus is almost completely protected from any possible injury. This point of view now is being modified. If one disease such as German measles and one drug such as Thalidomide can be harmful to the fetus, many others also may be injurious.

In Scientific Centers around the world a great deal of time is now devoted to the study of the embryo and fetus and the possible ways of preventing injury, arrest or stunting of its development. The most important period of pregnancy as far as danger to the embryo and fetus is concerned is the first three months and especially the first few weeks. Timing is important in determining the type of defect. Time, however, is not the only

factor, the type of noxious agent also influences the total effect on the fetus. German measles in early pregnancy tends to produce cataracts, deafness, congenital heart disease, and often cerebral palsy. Thalidomide on the other hand more often interferes with normal development of the limbs so that the infants after such drug therapy for the mother may be born without arms or legs or both. These signs and symptoms are characteristic enough so that a doctor working in the field when examining a child can often correctly name the cause.

Another cause of defects in infants is the Rh factor. About 15% of people are Rh negative. This refers to certain type of blood group factor. If the mother is Rh negative and the infant is positive, in some instances a reaction is set up between the mother and fetus something like an allergy. When this occurs the baby's red blood cells are destroyed, the infant becomes anemic, jaundiced and sometimes swollen. He may actually look waterlogged when he is born. There are methods to test the mother during pregnancy to determine if the Rh reaction is occuring. This test is known as a Titer. If the Titer rises, the physician in charge prepares to do a replacement transfusion of the baby immediately after birth. The replacement transfusion, as the name implies, differs from the usual blood transfusion in that the infant's blood is gradually removed and almost entirely replaced with other blood. In some cases, two or even three replacements are done. In the past few years a method has been developed whereby the infant is transfused while still in the uterus. This technique obviously is more difficult but it has proven successful in some instances.

There are other blood incompatabilities that may cause jaundice. The next most common one is known as ABO incompatability. This again refers to blood groups. When this occurs the jaundice is usually not as severe as that produced by the Rh factor. A negative Rh factor does not always create problems. Some Rh negative mothers have had six or eight normal babies without trouble. Usually the first baby is unaffected. After one baby has been jaundiced the subsequent ones as a rule are also jaundiced unless one of them happens to be Rh negative like the mother.

Between 6% and 7% of babies are delivered before the nine months of pregnancy have elapsed. These babies are called premature. With care, babies born two months prematurely, or weighing 2 lbs. and even less, have a good chance of surviving but they are more apt to have defects than the full term babies. Often the defects are relatively minor such as crossed eyes (strabismus), late walking and talking, or learning problems when they enter school. Sometimes they are mentally deficient or have cerebral palsy. In other instances they are born early because they have defects, rather than that the defects are caused by the prematurity.

Even when the well grown fetus arrives at term, that is, after a nine months pregnancy, the hazard of being born remains. It is important that he make a "soft landing". Like our astronauts, a safe landing is the most difficult part of the "orbiting". There are a great many things that can happen at the time of delivery that may cause injury to the baby. The baby who comes feet or breech first instead of head first is exposed to greater risk. When air strikes any part of the infant's body it stimulates him to take a breath. If then, his head is still undelivered he may get fluid in his lungs which will interfere with normal breathing. Sometimes the cord is around the neck so tightly that this interferes with delivery and the establishment of normal breathing. At other times the placenta (afterbirth) has been implanted entirely or partially over the opening of the uterus and excessive bleeding occurs when delivery begins. The baby may then be born in shock, pale and not breathing. Occasionally unexpected disproportion between the mother's pelvis and baby's head makes delivery difficult and traumatic. It may seem to a lay person that a Caesarian section would be the easiest way to deliver a baby. This is not true. Even in the hands of experts the incidence of complications with Caesarian sections is greater than with normal delivery.

After birth, the child's brain continues to grow rapidly and during this period of rapid growth it is more susceptible to harm than when fully grown. The more common causes of insult during the post-natal period (after birth) are such diseases as meningitis and encephalitis. Other injuries such as skull fracture, concussion, near drowning, and poisons all may leave lasting scars in the nervous system. Toxins or poisons from disease germs or excessive dehydration (drying out) from diarrhea likewise may cause irremedial harm.

The embryo or fetus or child who has had an insult to the growing brain, unless the injury is fatal, recuperates and continues to grow and develop. His development may be slower than normal; certain parts of the body may be more affected than others; some of the effects may take a number of years to become apparent.

NORMAL DEVELOPMENT

In order to understand the child who is different, it is important to know the normal pattern of growth and development. Many careful studies of growth have been made and are available to parents and to scientists in the field. The ones familiar to most of us are the works of Gesell and Spock.

While in the uterus the embryonic and fetal rate of growth has been estimated as follows:

At ten days half the size of an ordinary pinhead.
At three weeks about the size of an apple seed.
At four weeks about the size of a grape.
At five weeks about the size of a walnut.
At six weeks the size of a golf ball.
At two months the size of a hen's egg.

By five months the fetus weighs 1-1/4 lbs.
By six months 2-1/4 lbs.
By eight months 4-1/4 lbs.

The period after birth conveniently may be divided into (1) infancy up to 10 or 12 months; (2) the runabout period from one to three years; (3) the pre-school child; (4) the school child; and (5) the adolescent.

The most important factor in infancy is the stimulation received from the mother. The infant learns to know his mother by voice, touch, smile as during bathing and feeding. In this inter-relationship is established trust, confidence, satisfaction and love. These are extremely important months. If a child is born with a major defect this relationship may be altered in many ways. The parent of a defective child has many serious adjustments to make. The mother may blame herself; every thought, act, dream or wish she had during pregnancy recurs to haunt her. She may have doubts about her husband's or his family's reaction to her and to the baby. With this inner turmoil and sorrow she often forgets to smile, to talk or cuddle and fondle the baby. This can be one of the first hurts to the child. If the defect in the child is one of the major special senses such as vision or hearing, another learning experience is interfered with in the infant and this seriously affects the development of the child.

The next childhood period, from one to three years, is a time of rapid learning but the learning is self-centered. The infant creeps, rolls over, crawls, pulls up, cruises, walks and finally becomes dextrous both with legs and arms. He also learns to talk and normally becomes toilet trained by three years. These are normal maturational processes. The urge is born in the child. He does not have to be taught to walk or talk or feed himself. He is eager to do it. Oftentimes he is so eager and so independent that the mother worries about his nutrition because he will not let her help him with his meal. He loves

to explore the world around him. To him shut doors, closed drawers, stacked pans, covered containers represent a world of mysteries all of which he wants to test out. This world is as great and exciting to a two year old as a city in a foreign land is to a traveler. To view it only from a play pen or a high chair is as frustrating to him as it would be for you to be confined to a hotel room and to be forced to view the city from a window the first time you visit Paris. The child with a major handicap will miss some or many of the experiences that a normal child has. If his arms or legs or both are paralyzed or rigid or ataxic he cannot do the exploring.

The child from three to six has mastered himself to a degree that he can now walk, talk and feed himself and take care of his basic needs. He therefore begins to look around and get curious about the many things he sees. The questions that come to him are such as these: Why are there girls and boys? What is right or wrong? What is true and false? Ideas crowd in on the child at a tremendous rate during this period. If the child has not learned to talk or if his hearing is impaired or if he begins to compare himself with others and finds himself deformed or incapable how does he react and how does the parent react? These reactions are important in the developing child. This is the age that the child may ask about his own defect.

Then comes the period of five to six years. The child is eager to go to school. He cannot wait to start. What happens then to the child who cannot learn to read, who finds numbers a mystery? Whose hands won't obey his desires and the scribblings fly out of the teacher's planned pattern? These are the tragedies of the child with a defect.

The tragedies do not end with childhood. The period of adolescence must still be faced with all the heartaches of that age increased by a defective body.

GROWTH IN A CHILD WITH A DEFECT

Children with major defects, whether they are physical or mental, have the same inherent pattern of development. It may be slowed up in all areas or it may develop in a lop-sided manner. The child crawls before he walks. If he is paralyzed he may be very late in crawling and hence he will also be late in walking. He needs good control over certain movements before he can take on others. The infant babbles and makes sounds before he is able to make these sounds into words. His sentences at first are short. He may put only two words together. The child with certain types of cerebral palsy has great difficulty controlling the muscles of the face and tongue and mouth, the muscles that make speech possible. He drools, he cannot even control his saliva. Toilet training is usually the last of these skills to be acquired, hence the child who cannot walk and cannot talk is too preoccupied trying to master these skills to be concerned with toilet training. Time passes and the severely handicapped child may learn so slowly that he runs out of time.

The physical growth, however, may and usually does continue. Even though growth is at a slower rate than normal, the child gets big, heavy and increasingly difficult to handle. Time passes for parents, too. As they get older it becomes more difficult for them to meet the round-the-clock demands of the physically handicapped child. The parents have also had time to consider that their handicapped child may outlive them. Preparation for future care must therefore be considered.

What we have said applies to the child with a severe physical handicap, with or without mental retardation. There are children who are born without much physical handicap but who are severely mentally retarded. These children may pose an equally staggering problem to parents. Again, when they are little they go through the same developmental stages as the normal child but at a slower rate. They may and usually do acquire the walking skill fairly early. They may be able to feed themselves. Their speech, even though it is limited,

is present. Their deficiency is in judgment and discrimination. These children often are hyperactive. They may have to be watched almost continuously. They are often stubborn. They may be rough. They get angry, they do not obey and they have no caution so they cannot be trusted unattended. The problems with this type of child are different but no less difficult than with the physically handicapped child. For many of the physically and mentally handicapped children provision for some respite away from home becomes a necessity.

Although the child's own home is the best place for a handicapped child while he is little, after six or seven years he can adjust and sometimes be happier in a special school or in a boarding school, if necessary, just as many normal children attend boarding schools during childhood. He may be happier in a situation where the training and other activities including play are planned around a group of children of about equal ability. It may be lonelier for him at home where he is trying to compete with normal brothers and sisters and normal children in the neighborhood.

LABORATORY TESTS

There are certain medical tests about which parents often ask questions. Many handicapped children either are retarded mentally or have learning problems. Tests concerned with the study of the nervous system are performed frequently. A simple x-ray of the skull gives us some information. It may reveal unusual thinness or thickness of the bones. It shows whether or not soft parts called fontanels and the seams or sutures are the proper size, in the usual position, or closing in infants too fast or too slowly, or are abnormal in any other way. Sometimes an x-ray picture shows a calcified blood clot or a tumor with calcification in it. There are a few diseases that also cause spots of calcification within the brain itself. Calcification means deposits of calcium as in bone. X-rays do not penetrate bone readily and such deposits are poorly visible.

The fact that x-ray does not readily penetrate bones also limits the use of plain x-rays in the diagnosis of disease of the nervous system. A pneumoencephalogram gives more information. The word "pneumo" means air and "cephalogram" refers to the brain. The pneumoencephalogram carries the study of x-ray of the brain a step further. This test in children is done under anesthesia. The brain is not solid. It has several cavities called ventricles that normally are filled with spinal fluid. This makes it possible to remove some of the fluid and study it in many ways. Tapping is not difficult and not particularly painful, not more so than any shot with a fair sized needle. The spinal tap needle is hollow. After the fluid is withdrawn, by letting it drip out through the hollow needle inserted into the lower spine, the needle is left in place and a syringe of air is injected. The amount of air depends on how much fluid is removed and on the size of the child. X-rays are then made of the skull and the air shows up the size of the ventricle, any thinning of the brain or any lopsidedness such as a large ventricle on one side and the small one on the other. This test, therefore, gives us much more information than the plain x-ray. It can be compared to a study of the stomach and intestines by using Barium as a contrast medium. The time to use a pneumoencephalogram must be left to the judgment of the physician.

Another test occasionally used is an arteriogram. As the word implies, this test gives information about the blood vessels or arteries of the brain. It is used particularly if a tumor or abscess or scar is suspected as a cause of the child's problems. Sometimes a child with a large red birthmark on the face or scalp may have neurological findings. An arteriogram is then valuable in determining whether or not there is an abnormality of the blood vessels or of the circulation of the brain.

The electroencephalogram or brain wave, EEG as it is commonly called, is not as valuable as parents and as many teachers think. It is not nearly as valuable in a study of the brain as the electrocardiogram is in the study of the heart. The test must be taken over skin

and bone and obviously cannot give very detailed information about the inner workings of an organ as complicated as the brain. The EEG, however, is of particular value in detecting convulsions and different types of epilepsy. The important thing to learn is that a normal brain wave does not mean "there is nothing wrong with the brain". There may still be neurological problems even with a normal brain wave. An abnormal brain wave may simply represent immaturity or delay in development and does not necessarily mean serious pathology nor does it necessarily mean that anything can be done about it.

The chromosome count, another laboratory procedure, has been mentioned earlier. It is not done often because many of the diseases due to an abnormal chromosomal count can be recognized without the test. It is a difficult and an expensive procedure - only the larger laboratories are equipped to do this test. It is used in uncertain or suspect cases and sometimes to detect carriers.

Blood chemistries are used fairly extensively. The PKU (phenylketonuria) test is now required by State law to be done on all newborns. The test can be done either on the urine or the blood. The urine test is less reliable so the blood test known as the Guthrie test is usually done.

There are many other chemical tests that are made to either diagnose or rule out certain conditions. Among these tests may be mentioned blood sugar levels, calcium and phosphorus levels, certain amino acids, and so forth.

The thyroid test is sometimes run to be sure that the child has an adequate amount of thyroid secretion. Cretins are born deficient in thyroid secretion.

THE PSYCHOLOGICAL TEST

The psychological test is done on almost every child at our Center. This test is not done to get a fixed I.Q. or intelligence quotient. It is an extensive test sometimes, especially on young children, requiring two or three visits. The aim is to get a profile of the child's aptitude and ability much like an adult seeking a job may take a series of aptitude tests. We have many well performed studies on normal children. Psychological testing is a comparison in many areas of a particular child with the normal children of the same age. The I.Q. is the ratio between the real age of the child and the age he tests at. Mothers who have more than one child often describe the child's performance quite accurately. If the question is asked "How old does your child seem to you?", it is surprising how near to the actual test score the mother's own estimate comes. She will often mention the areas in which he is different; for instance, the mother may say of her four year old child, "He is about as able as his two year old sister, but he doesn't talk as well as she does". The test may show over-all I.Q. at about 50 but his verbal performance lower.

Even in the pre-school child a careful psychological test can anticipate future learning problems such as difficulty in learning to read, in dealing with numbers, or in writing. It is important to have such information. A child with these difficulties should be helped in advance and spared the humiliation and frustration of failure and the emotional consequences of such experiences.

SPEECH

The speech evaluation is also important. Children start talking at different ages just as they do walking. Age at which speech develops is not necessarily related to intelligence. Some quite seriously retarded children talk a great deal - they may not say much, but they can be very verbal. Some children with normal, even superior intelligence, may have very limited speech even at 2-1/2 or 3 years. Any question about speech that arises either with parents or relatives or teachers should be investigated. It is especially important to

test for hearing. Even in a child there may be degrees of hearing loss. A child may not hear high frequency sounds or low tones. If he is born with such a defect he naturally will not learn to use or imitate sounds of which he has no knowledge.

SEX DIFFERENCES AND LEARNING PROBLEMS

Problems in learning in early childhood are more common in boys than in girls. The reason for this is not generally known or recognized. Girls mature earlier than boys. This is an accepted fact, the legal age for men being 21 years and for girls 18 years. It is also apparent in early adolescence that girls are maturing or growing up one to three years earlier than the boys. Curiously this knowledge has not been generally applied in the schools at the First Grade level. Reading readiness is as much a reality as walking readiness. An infant cannot be taught to walk until he has matured to a certain state. The child will not be able to read until his maturation for reading has been attained.

SOME CHARACTERISTICS OF CHILDREN WITH DEFECTS

The word "retardation" means different things to different people. To some parents it means idiocy or feeblemindedness. Some parents think it means the child will never grow beyond what he is now. The word more accurately means "behind for his age". There are, therefore, degrees of retardation. Some children are behind their peers because they have been seriously ill. Others may have been prematurely born and have been slow in getting started. Some have been deprived because of poverty and discrimination. Others are actually mentally deficient.

If a child is moderately or severely handicapped his daily care may call for special techniques. A child with cerebral palsy usually starts out in life crying a great deal. Almost all infants with a defect of the brain are restless babies. They are wakeful and cry a lot. Parents are tired when they lose a great deal of sleep. These infants seem to be resistant to most drugs that are used in quieting children. These babies are also poor feeders. They have difficulty sucking on the bottle, they choke easily on the food. It requires patience and time on the part of the mother. At the same time children who are small for their age do not need as much food as big active youngsters and mothers should not be too anxious over the amounts a child eats.

Many of the children with a neurological deficit are constipated. This, in part, may be due to a poor appetite but also it is partially caused by the rather spastic muscles of the intestines.

Many handicapped children have eyes that cross and often it is necessary to have them corrected by surgery. Sometimes one or both tear ducts are closed, the eyes water a great deal and may get infected. This defect, too, is fairly easily corrected.

Infants and young children with handicaps seem to regulate their heat mechanism as well as the normal. They do not need to be dressed more warmly than other children. They are usually more susceptible to infections of all kinds. They often snort when they breathe and may sound very noisy. These sounds are of no real importance.

It is important that the children be immunized as are normal children. If surgery of any kind is necessary they tolerate the anesthetic and the surgery very well, but both the anesthetist and the surgeon should be selected with care as even physicians who are not accustomed to these youngsters are often somewhat hesitant about undertaking surgical procedures on them. It often is necessary to give an anesthetic in order to do a complete examination or a repair job on the teeth.

Children with neurological deficits are somewhat more prone to convulsions or seizures,

These may be "grand mal", which means the child has a real shaking fit or they may be "petit mal" which implies that the seizure is small and short, that is, the child stares into space for a second only. Often the parents look upon these latter attacks as "day-dreaming".

LIFE EXPECTANCY

Parents who have a severely handicapped child may ask (but more often they only think about it themselves): "How long will he live?, or if he outlives me, what will happen to him?" These are normal thoughts and such questions should be asked even though a reliable answer can seldom be given. Those who make positive predictions are usually inexperienced and uninformed. As has been stated earlier, most severely handicapped children are stunted in their overall development. They are fragile and susceptible to infection. In contrast, well designed human beings like Winston Churchill and Herbert Hoover are strong physically, mentally and emotionally and live long. It can be said, therefore, that the life span of severely handicapped individuals is shortened but how much it is shortened is impossible to state. In a few individuals who suffer from certain specific diseases, the survival time is predictable. An example of such is Tay-Sachs disease which is a hereditary progressive condition which, if present at birth usually the child does not survive beyond three or four years. Diseases of this nature are rare. Most retarded and defective individuals may live to adult life and a few into old age.

COMMUNITY FACILITIES

Parents often want to know what provisions have communities made to meet the needs of the handicapped. There are many provisions and more are being added every year. All children have the same basic needs. These needs are a home, medical care, education and training up to his ability, recreation and religion. These will be taken up in order.

Home

His own home is the best place for the young child, if this is at all possible. He needs tender loving care as any child does. He needs to belong. He needs protection but also stimulation. He needs to develop confidence in himself and he needs to feel he is a person who is worthwhile and who is wanted. If he has no home of his own then a foster home is the best substitute.

When the child reaches the age of six to ten other provisions than home may be necessary and may be desirable. If the child is severely physically handicapped and is also large, he may add to his normal weight that of heavy braces. Such a youngster may need more nursing care than the mother alone can give. There should be boarding schools for these children where they can get help, supervision, education and training but like other boarding students they can have weekends and vacations at home.

The retarded hyperactive child, and even the hyperactive child who has normal or superior intelligence, may need a boarding school. Unfortunately at present very poor provision is made for such boarding care at a price most parents can meet.

The child past adolescence and the adult who is too handicapped to take care of himself needs some type of permanent help. The State provides for this and an increasing number of churches are also making provision for permanent care for dependent individuals.

Medical Care

At present the medical care is most adequately provided; no child needs to go without the necessary medical care. The cost may be met through insurance, through hospital clinics

through Crippled Children's Services and the like. The child with a handicap should receive the same kind of medical care as the normal infant or child. He can be immunized against whooping cough, diptheria, tetanus, polio, smallpox and measles. He can take antibiotics when he needs them. He tolerates surgery.

Education and Training

The public schools have made many provisions for the education of handicapped individuals, and they are continually adding new classes to meet needs. In California there are special classes from three years and on for the deaf, blind, cerebral palsy, and other motor handicaps. There are also special classes for the mildly retarded, the severely retarded, and the so-called educationally handicapped. These latter are children who have such problems as reading (dyslexia), number difficulties, (discalculus), and speech problems (aphasia). These are good programs but they do not meet the needs of all the handicapped children. Private agencies, largely started and sponsored by parents are not only trying to fill the gaps but are also working to get the public agencies to expand their services. Through such private sponsorship nursery schools and pre-nursery schools are being provided for the children with cerebral palsy, deafness, retardation and emotional disturbance. The latest device that is now being developed is that of Day Care Services for children who are so severely handicapped, either physically or mentally, or both that they cannot profit from any of the above educational facilities.

For the older children and young adults special workshops are provided. There is also a constant campaign to get industry to hire the handicapped.

Recreation

Children who are either physically or mentally handicapped need to laugh and play and have fun. Often grownups act as if the only thing a child needs is to learn to read and write. Tremendous effort goes into teaching the retarded children letters and numbers. Young children learn best by play. They play out all roles before they read or talk about them. Childhood conversation sounds like this: "You be papa. I'll be mama. Susan is the littlest, she will be the baby". Then the little mama may be using your words to talk to Susan like, "You bad, bad girl. I'll spank you if you do it again", or if you are not that kind of a mama you will still hear yourself quoted. A child who is retarded, that is, if at 10 years he is developed to only a three year level, he still learns like a three year old. He should be helped, therefore, to learn at this level through play. Three year old girls like to set the table, maybe they try to dust; boys may like to feed the pet, rake the leaves and play at work in other ways. Special recreation for the level of performance should be provided for all children who are unable to join completely into the activities of the normal children in the community.

Religious Training and Enjoyment

The Churches have pioneered many services. There are Church supported hospitals, colleges, nurses' training schools, old folks homes, and so on. Very little, however, has been done for the physically handicapped, or the retarded child. This is a curious phenomenum that is hard to explain. The pastors say that these problems are seldom brought to them and the parents often say, "No, I have not talked this over with my pastor". This lack is being corrected. In California the Good Shepherd at Terra Bella is sponsored by the Lutheran Church. In Northern California a similar sponsorship has been started by "The Helpers of the Holy Innocents" in the Catholic Church. Both of these are non-denominational.

SPECIALISTS

The severely handicapped child may have to see many specialists. Not all parents understand the differences between specialists. It may be well therefore to explain briefly the functions of different doctors.

A pediatrician is a child specialist, a doctor who limits his care to the young, usually up to 18 or 21 years of age.

An orthopedist deals mostly with bone deformities such as club feet, broken bones, crooked backs. He does surgery in this field as well as medical care, bracing, providing crutches, etc.

The neurologist specializes in diseases of the brain and the nerves extending out from the brain and spinal cord. In addition to making a diagnosis of these difficult problems, he deals particularly with convulsions, epilepsy, and other diseases of the nerves and brain. He is not a surgeon.

The neurosurgeon does surgery on the brain and spinal cord when it must be done. There is always some overlapping of work in these specialties.

The ophthalmologist is concerned with abnormalities of the eye.

The ear, nose and throat doctor, otolaryngolist, as the name implies, handles the problems that deal with these organs.

In the more recent years a physiatrist, or M.D. of physical medicine, has been put in charge of programs dealing with rehabilitation. In childhood habilitation describes more accurately what is needed than the word rehabilitation. If you redo something it means you have done it before. Since handicapped children usually are learning it for the first time, the word should be "habilitation". The physiatrist, therefore, helps the child to gain control of muscles. The physiotherapist and the occupational therapist carry out the orders of the doctor in charge of habilitation. The physiatrist is usually trained in orthopedics.

The psychiatrist is a doctor with special training in the area of emotions or feelings. Unfortunately many people think of a psychiatrist as being concerned only with people who are psychotic or, in lay terms, "crazy". This concept is incorrect and works against getting help often when it is sorely needed. The psychiatrist is well trained in the field of emotions. He understands actions and reactions and by diagnosing and interpreting these inter-reactions many problems and worries can be solved. To give a simple example, if a child of four or five years sees his home burn down and the parents and siblings are very frightened and upset by it, then, in later years, (and usually for the rest of his life) he will react with fear when he sees an open fire. Or, if lightning should strike his home the sight or sound of lightning and thunder may bring a reaction of fear and distress in years to come. In a similar way any unhappy or frightening or lonely or anxious experience of early childhood may be triggered off later in life by events which suggest or recall these early experiences, even though the individual himself has forgotten them.

The psychologist works in this same field but he is not a physician. He usually works under and with a psychiatrist and other doctors in the field. He is trained especially in evaluating the child's intellectual performance.

The physiotherapist works under the direction of an orthopedist or a physiatrist to help overcome deformities and to strengthen muscles.

The occupational therapist assists in the above work by helping the child gain skills in many areas such as dressing and feeding himself, buttoning, tying, and the like.

The speech therapist and audiologist is trained in testing hearing and in analyzing speech and in helping the child to attain normal speech.

A social worker assists with home and community problems. She is concerned with talking to parents, with contacting schools, nursing homes and other agencies in the community that may be of help to the parent with a handicapped child.

Educators are teachers and assist in helping the child gain the necessary experience to grow to be ready for the next school or learning situation.

The public health nurse makes home visits and assists the physician in many ways to see that the doctor's orders are carried out. She can also interpret the home situation to the physician.

Each specialist plays his own role in the planning, diagnosis, treatment and training of the child.

REACTIONS TO THE HANDICAPPED CHILD

When a child is greatly handicapped either physically or mentally or both it is noticeable. People will not only stare but they will ask questions and often make suggestions. It is best to meet this situation straightforward and honestly; (even a little humor helps). Interpret the child to the relatives and neighbors, give them the diagnosis as you understand it. Other children will be kind and helpful if they are told what is wrong. We are all a little awed by the unknown and when we are, we shy away. When in doubt ask your doctor or social worker how to handle the situation.

Some parents like to read about the child's condition. There are many books written on almost every subject. There are also journals on almost every subject. It is usually best to ask your doctor or teacher or another member of the staff working with your child about the subject matter. There are journals published by such organizations as Cerebral Palsy, Muscular Dystrophy, Aid to Retarded Children, and many others. Almost every disease has its own organization. You may want to participate in some way with the appropriate organization. By banding together parents have accomplished a great deal to get better services for their children.

You are not alone with a problem.

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A MANUAL for Parents of Handicapped

THE HEALTH-RELATED PROFESSIONS AND PEDIATRICIANS

By

1970

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There is pressure now in Pediatric circles to add another health-related profession--a doctor's assistant. An astute volunteer on a hospital ward made this remark, "The more workers in the field, the less care the patient gets". In 1969, the New York Academy of Science published the papers of the Symposium on "Education in the Health-Related Professions". One purpose of the symposium was to emphasize the role of the various workers in the health field in augmenting the distribution of health services due to the medical manpower shortage. Special emphasis was placed on higher education and advanced degrees in these professions.

After many years experience working with health teams in rendering services to the handicapped, it seems appropriate to emphasize both the advantages and the problems posed by group work. Let's leave the best to the last and start with the problems.

One of the problems is the cost -- the larger the staff the higher the price. Money for service is difficult to come by. For many years both public and private funds have been funneled into the Universities for research projects. It has been practically impossible to get money for service programs. The Crippled Childrens Services will finance diagnostic studies but not continuing therapy. Fee for services provides only a token sum and only a rare family can meet the costs privately, of a long term medical problem.

The cost is only one of the problems. Another one is "degrees". By the time a professional person has acquired an M.A. or a Ph.D. degree he or she wants an administrative or consultant position. Daily floor work with the patients is too menial for them. This latter, however, is the real need and this work is difficult. In addition to therapy it often means wiping runny noses, changing dirty diapers, soothing weeping youngsters and consoling distraught nothers.

A third problem is to find a group of people who are congenial and who can work together without friction. A psychiatrist once said to me, "It takes ten years to mold a successful team". Usually it need not take that long. If a new service is contemplated in a hospital or clinic setting it is well to start with a few people already working in the field and acquainted with each other. As additional personnel is added, a new member should be interviewed by several members of the group. It is also judicious if possible to accept new workers on a trial basis for a period of two or three months.

It is important also for the members of a team to define their roles and adhere to them. In dealing with patients over a long period of time, it is not unusual to have some who will try to manipulate the staff or to play one member of a team against another. A young worker may fall for this and tend to take over the role of a fellow worker. This may occur in counseling, in speech therapy, or in occupational therapy. The worker must be able to say, if faced with this problem, "Let's take this question to the right party".

A frequent problem, especially at the Ph.D., level is to find the workers talking to each other instead of to the patients. Conferences of staff

members is important but one or two hours a week is enough. At these meetings the focus must be on the patient and his needs. Talking alone does not get the nitty-gritty work done. It is hard work with patients that needs to be accomplished.

When the above problems are under control team work can be very rewarding -- even exciting. The patients make progress, the mothers relax and enjoy each other, and the staff profits from a satisfactory feedback. When working with the permanently handicapped or a long term illness, progress is slow. The youth of today want instant answers and probably the most difficult lesson for a new worker to learn is patience. To counteract tedium the leader needs to understand the staff, to recognize fatigue and discouragement. New ideas and insights are important. These can be introduced periodically by having community leaders attend the weekly sessions and participate in the discussions. The staff members should be encouraged also to attend pertinent medical rounds, to participate in local, state and national meetings of their specialty, and to have on occasion a leave of absence for futher study or training. They should be encouraged to engage in investigative work or original studies and present or publish their findings.

Physicians, until recently, have been trained largely to be solo performers. They have concentrated on disease entities. Pediatricians have been so successful, both in the prevention and care of many childhood diseases, that to some the glamor of practice is gone. This need not be so. The new challenges are multiplying rapidly. The prevention and cure of social and emotional ills confront us. The pollution of our planet, the population explosion, drug abuse, and the prevention of the birth of

defective children are all as challenging as were diphtheria, meningococcus disease or the black plague in their day.

Pediatricians, more than any other medical specialists are prepared to meet these challenges. The Academy of Pediatrics since its origin has had tentacles out in every direction, it has representatives to all other organizations, dealing with children or with health problems.

Pediatricians can show the way of how to use the health professionals to extend the arm of health care to all the children in our nation and then in the world.

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The HEALTH-
RELATED Program

TEN YEARS EXPERIENCE WITH CEREBRAL PALSY AND RELATED DISEASES

With ten years of experience behind us it seems timely to review changes that have taken place in the field of Cerebral Palsy in the past decade.

The staff at Children's Hospital has been exceptionally stable. Mrs. Wilda M. Flower, Mr. E. Walton Kirk, Mrs. Jane Sledge, Mrs. Gertrude Saxton, Dr. Lucille Eising, Dr. Elsa Cabos and Dr. H. E. Thelander have been with the program either from the start or shortly after its inception. The medical consultants are essentially the same as the original group. The 1961-1962 report shows that many of the original patients are still being followed. Nearly 1000 patients have been seen which should give us a fair sampling of the problems encountered among neurologically handicapped children and their families. Furthermore, the staff members have participated actively in many and varied organizations dealing with the handicapped at local, State and National levels. Some members have visited centers abroad and many foreign visitors have been our guests. These exchanges have been very helpful in evaluating the validity of our own observations and experiences.

In general a medical problem is attacked by Research, Education and Service. The AMA Journal of June 30, 1962, has an illustration showing a patient toppling off a tripod due to the broken short service leg, the other two legs being Research and Education. The article gives the following statistics: In 1940 the Government expenditures for research were 53 million or about 7% of the total public and private medical research support. In 1961 this amounted to 496 million or 56% of the total. It is predicted that by 1970 it will reach 1.6 billion. The Institute of Neurological Diseases and Blindness of the National Institute of Health is spending millions on the causes, prevention and cure of neurological diseases in children. NIH grants include the training of personnel in the fields involved. The field of Research has obviously expanded tremendously.

The field of Education, both medical and Ancillary, has also expanded greatly. The July 1962 number of the Medical Dates Bulletin of the CMA has already listed 82 medical meetings for the months ahead. Several of the meetings pertain to subjects associated with Cerebral Palsy and its related dysfunctions. It has become almost impossible to schedule meetings without one or more conflicts. This shows an impressive interest but also poses problems. With such a large and varied number of meetings to choose from the doctors or other participants naturally select those in which they are most interested. This means that meetings centered about a given disease entity attracts primarily those already interested. It does not reach out to the other fields of medicine where information is most needed. At a recent meeting in New York, the directors of five Development Centers from New York to California were of the opinion that to succeed in putting across the already available information on causes, prevention, and cures of neurological diseases it was essential to get the subject as well as the result of our research in some form, under some type of title, into the programs of Obstetricians, Internists, Surgeons, Anaesthetists, Radiologists, Pharmacologists, and Pediatricians. Furthermore, the subject of Child Development and the hazards peculiar to a rapidly growing nervous system should be taught to all medical students at the undergraduate level. This would cost nothing extra. Furthermore, since fees for refresher courses are deductable items on the income tax, refresher courses could be made self supporting. As a matter of fact, transportation and honorariums have become a means of vacation and travel for many popular speakers. This becomes an unwarranted luxury unless the patients benefit from the application of this latest knowledge.

Service to the patient has not followed the expansion pattern of Research and Education. This fact warrants serious consideration. The needs of the Cerebral Palsy patients in San Francisco are not met. This is particularly distressing since the number of Cerebral Palsy patients is not great. San Francisco is a unique county in being entirely urban and its suburbs are almost totally in other counties. The population of San Francisco has actually decreased while the neighboring counties have increased. San Francisco, with

two Medical schools until recently; with a Teachers College, and State and local institutions should have developed an ideal or model program that could have been an inspiration to growing and developing areas. With the tax funds and the private funds collected for the handicapped and available to this city, lack of money cannot be the sole cause of this situation.

Basic to adequate services are: (1) Transportation. An excellent program is of no help to the patient who has no means of getting there. Consider a mother without a car, with limited finances, and possibly with 2 or 3 preschool children, attempting to bring a heavy, severely handicapped child across the city to a Day Care Center; or a mother who is employed trying to get even a single child transported and picked up. These are daily realities. (2) Ancillary services such as Day Care, training programs for the handicapped and recreation should be without cost to the handicapped as they are to the normal children. (3) Screening children out of existing programs without making any provision for, or taking any responsibility for children so barred from help is a deplorable situation in our communities and should not be tolerated. There should be a "home base or bases" to which such patients could be referred by the agency that turns them away and another means of help found. Existing institutions could be more elastic and accommodate the relatively few "hard to place" cases than they are at present. Unfortunately many programs start by defining what they will do and then close their eyes completely to the needs of anyone outside the limits set.

The unmet needs of the neurologically handicapped children in San Francisco and the methods by which they could be met are outlined below:

1. THE SEVERELY PHYSICALLY AND MENTALLY HANDICAPPED. (Confined to bed.) This group is not large. There are less than 100 children on the waiting list for Sonoma. Of these 100 probably not more than 20 would fall into the above category. There are probably another 20 or 25 in the same category but application to Sonoma has not been filed. From personal contact with many of these parents we know that they would utilize hospital beds intermittently if these were available and "commitment" was not demanded. The suggestion has therefore been made that the State or Local Mental Health Division or Sonoma (whoever is responsible) contract with the Hospitals of the city to hospitalize these patients in the regular Departments of Pediatrics until a transfer to a State Institution can be made. Since these patients have already been carefully diagnosed, hospitalization could be on a minimal care basis. Pediatric Departments tend to have an occupancy of 50% or only slightly over, beds and staff therefore are available. Although the number of these patients is not great, the hardship to the families is disproportionately grave.

2. DAY CARE AND OCCASIONAL BOARDING. (Patients screened out of all educational and training institutions.) If Day Care services were free and transportation provided there might eventually be 75 or possibly 100 children so cared for in the City. The exact number could not be determined without a trial. There are some parents who prefer to take care of the children at home. There are other children whose training status can only be determined by trial. There are a few children in existing facilities that might be more suitably taken care of in a Day Care situation. If provision were made now for 20 or 25 children it might take some time to fill the quota. It would be prudent therefore to start with a relatively small activity and provide for expansion if and when this becomes necessary. This is again a problem that concerns a relatively small group but one that poses disproportionately grave situations.

3. THE MULTIPLY HANDICAPPED. The past year Sunshine School provided two additional classes; one for the blind-neurologically handicapped and one for the hyperkinetic neurologically handicapped. There were ten children thus accommodated. We hope

these classes will be continued. One of our children who is deaf and hyperkinetic was sent to Napa State Hospital for lack of other residential facilities. A great deal of work by several agencies was done on this family, all of whom recognized that Napa State was not the appropriate facility but it was the only one available.

4. UNDER 8 YEARS, POINT TWO AMBULATORY CHILDREN. A class for 10 or 12 children between the ages of 5 and 8 years at Louise Lombard School would take care of a group of children not accepted in Sunshine School because the degree of their motor handicap in their opinion does not justify admittance. The Point II child is easily diagnosed before 8 years of age. It is well recognized that early training for these children is extremely important. The parents also need guidance in home training before 8 years of age.

5. AID TO RETARDED CHILDREN NURSERY SCHOOL. This facility accommodates some of our children who have become ambulat ry and who are not eligible for Sunshine. More parents could use it if transportation were p· ·vided and if mother participation were not required. Working mothers and some others cannot always arrange for participation.

6. APHASIA AND DYSLEXIA. Children with major or minor physical handicap but with aphasia or dyslexia pose a serious problem in this area of the country. The State Cerebral Palsy School with generous tax support remains a diagnostic and evaluation center and is of very limited value as an educational institution. A child with aphasia needs years, not weeks or months, of education. With the expansion of diagnostic centers throughout the State the function of this institution should be re-evaluated. The function of the Deaf and Blind State Schools should also be re-evaluated. At present they will not take neurologically handicapped deaf or blind children. Inasmuch as provisions for the education of the "normal" blind and deaf have expanded the question might well be asked "Do we need residential schools for "normal" blind and deaf children?" If some of these children come from remote or isolated areas, could they not board in a normal home and attend school in a nearby urban place? It seems to us that these institutions might need to change with the times and provide more urgent services such as long term training for the aphasic, the severe dyslexic, the blind and deaf neurologically handicapped and the adolescent severe cerebral palsy. Stanford Medical School is launching a project for aphasic children; being in a medical school it will probably be able to give only limited service.

7. PREPARATION FOR USEFUL LIVING. Our experience indicates that our schools are too academically oriented. The pupil finds himself at 16 or 18 years of age out of school and without any concept what he is to do next. This is late to turn to rehabilitation. Actually for the child with a permanent physical handicap orientation toward a realistic work goal should start at 12 years of age and the schools should be so geared. From the experience in sheltered work shops IQ's by standard testing are not the total indication of ability to perform in a work program. Since most of the children are in need of practical rather than academic training, special education should be directed toward work performance when the children reach the age of 12 or 13 years.

The professional concept of Cerebral Palsy has changed in the past decade. This is aptly illustrated by the British who in January of this year changed the name of the "Cerebral Palsy Bulletin" to "Human Development and Childhood Neurology". President Kennedy in defining Mental Retardation to his committee for the study of the subject made that definition so broad that for practical purposes it covers 75% or more of the Cerebral Palsy cases. At the local level, Dr. Kimmicks committee on Mental Retardation accepted President Kennedy's definition as a working definition. The preliminary tenor of the above two groups (President's committee - Short Doyle Funds) is toward SERVICE to the patient. We can only hope they are not deflected from this course.

Also in the past decade there has been at least two new organizations in the field, one for the Neurologically Handicapped and one for the Multiply Handicapped. Groups such as these are formed because services are not available to the children. If the existing organizations supplied the services new parent groups would not have been formed. On the other hand there has been a tendency also for parents of handicapped children to unite. "Concern" is in part such a group. At the local level there has been at least two joint meetings of parent groups, and a third is contemplated. Parents are not the only ones seriously concerned about the lack of services, donors increasingly are disturbed by the multiple demands for their charity dollars. To both parents and donors the super structure seems to grow and expand becoming more complicated and more remote from and with less services to the patient.

The cartoon cited earlier pictured the patient falling off the tripod due to the short broken service leg, it is axiomatic, however, if services break down the super structure will topple eventually too.

A great deal has been learned as a staff working intimately with patients and families, some now for ten years, as well as by exchanging experiences with others doing the same type of work. There is general agreement as to what is needed. Surveys, demonstrations, and pilot studies are passe in the field of the neurologically handicapped. Temporary grants are a delusion; steady continued endeavor is needed. Programming for these children is neither extremely difficult nor unduly expensive. The focus must be on the patient and his immediate family; and then the skills of the different staff members must be effectively utilized. This means recognizing and respecting the roles of every member of the program. It is an Orchestral rather than a team approach.

Recognition of human worth needs to be re-emphasized. It has happened time and again when parents have a satisfying diagnosis and interpretation of their problem and some support in dealing with it, they in turn interpret it to their neighbors, the church, and the school. Many families have found ready help and cooperation all around them. When people begin to help people there is not much need for a complicated and expensive super-structure.

Although progress has been slow and often discouraging the past ten years, the momentum appears to be increasing and the next ten years should show far greater accomplishments.

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With the assistance of:

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CONSEQUENCES OF PERINATAL INJURY TO FETUSES WITH EARLY NORMAL DEVELOPMENT

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CONSEQUENCES OF PERINATAL INJURY TO FETUSES WITH EARLY NORMAL DEVELOPMENT

Introduction

In a previous study[1] the possible causes and contributing factors to congenital malformations were explored; the present study is designed to review clinical abnormalities resulting from injuries occuring during the perinatal period to normal fetuses. It is not always easy to differentiate between birth defects arising during organogenesis and those occuring secondary to neonatal adverse factors. Hemiplegia, usually due to birth trauma, has been seen as a hereditary condition in our series; and microcephally[2] has resulted from neonatal meningitis in a normal newborn; then there is always the possibility that an unrecognized anomaly predisposes the infant to injury at the time of delivery.

Although the basic organization of the body is programmed into the conceptus and laid down in the first few months of intra-uterine life, growth is not complete at birth and development may be affected by adverse factors in the perinatal and newborn periods.

The approach to the study will be 1) a review of the development of the brain and central nervous system, the anatomy of the brain and the localization of functions as understood at present; 2) a review of the pathology of brains in still-births and neonatal deaths of presumably normally developed fetuses. 3) In certain well documented cases, an attempt will be made to correlate birth history and subsequent clinical symptoms in the light of the above studies.

Of special interest is the preservation of normal or even superior intelligence in individuals with severe physical impairment from birth injury to the brain.

DEVELOPMENT OF THE CENTRAL NERVOUS SYSTEM

During the period of 9 to 11 days which elapses between fertilization and implantation of the ovum,[3] the conceptus receives its nourishment from the yolk sac, from nutrients in the fallopian tube, and from secretions in the uterus. After implantation, placental formation, and the establishment of umbilical circulation, nutrients are obtained from the blood. It is important to appreciate not only the availability of these nutrients but also the minute size of the conceptus and embryo during the early weeks of pregnancy.[4] At 4 weeks, the crown-rump (C-R) length is 5 mm (0.2 inch). The neural tube is laid down between 3 and 4 weeks, but at 5 weeks the length of the embryo is only 8 mm (0.3 inch). By 8 weeks or 2 months it is 23 mm (0.9 inch) and weighs 1 gram. By this time the frontal area of the embryo has enlarged and differentiation is apparent. At 3 months the length of the embryo is 56 mm (2.2 inches), and a month later it is 112 mm (4.5 inches) and beginning to look human.

At the end of the first and during the second month, the choroid plexus[4] begins to form and secretes large amounts of protein into the spinal fluid. At this stage, the spinal fluid cavity is closed. The concentration of protein in the fetal spinal fluid is roughly 20 times that in post-fetal life, and also serves as a nutrient to the developing embryo. Only after 3 months of extra-uterine life does the choroid plexus and the spinal fluid develop the characteristics of adult life.

The blood vessels of the embryo begin to penetrate the brain tissue during

the second intrauterine month, and largely signifies the end of the role of any other nutrient supply. As the vascular system of the fetus takes over, it becomes increasingly important in supplying oxygen, glucose, and other nutrients. The ventricular system, however, remains closed until the fifth or sixth month, when the foramina open and spinal fluid circulation becomes possible. The spinal fluid then bathes the surfaces of the brain. The newly-formed brain cells in the periventricular matrix migrate toward the ingrowing blood vessels.[5,6]

The nucleated red cells of the blood lose their nuclei and become better adapted to transfer oxygen. These nucleated cells use oxygen for their own needs, and embryonic hemoglobin absorbs oxygen avidly. Furthermore, there are other distinguishing features in the morphological structure of the cerebral capillaries. They do not contain connective tissue elements in their walls. Also, it is generally believed that the cerebral capillaries do not contain elastic membranes, and argyll fibers appear only when the fetus matures. The elastic coats appear with aging and then not in all the brain areas. Dye tests which stain other tissues of the body do not enter the central nervous system; the capillary walls are not permeable to them. This is very significant when considering the blood brain barrier. In asphyxia, the capillaries of the brain dilate whereas in most other tissues they contract. Epinephrine, likewise, reacts differently on the brain vessels than in other tissues. Clearly, the nerve cells are dependent on an uninterrupted flow of oxygen. Klosovoski[6 & 7] notes that a slight and short-lasting asphyxia stimulates growth of new capillaries; even the products of cerebral tissue degeneration following trauma, have a growth-stimulating effect.

The brain cells need an uninterrupted flow of both oxygen and nourishing substance. The metabolism of fats, carbohydrates and proteins is extremely

rapid[7] and the speed of the blood flow also rapidly discards metabolites. The oxygen uptake of the central nervous system is high in respect to other tissues, and amounts to 20% of the uptake of the body. The brain is almost equally dependent on glucose, the major energy source of the brain, and consumes it at the rate of 5.4 mg/100 gm/minute.[8]

The growth of the brain under varying circumstances is significant. Minkowski et al[9] noted that the development of the hemispheres generally is steady and even. This appears even when there is great variation in the weight of the infants as presented in his graphs to show correlation of the weight of the brain and the weight of infants with the length of gestation and the varying weight gains of the fetus. In discrepancies of weight, as for instance in the dysmature or very small-for-age infants, the brain seems to suffer very little in morpholofy and weight, even though the body or total weight is far below the median line. Similarly, in diabetic mothers, where the infant may be unusually heavy, the brain appears unaffected in development and maturation.

Baker et al[10] in their impressive study of fatal cases of bulbar poliomyelitis note that an individual might have an alteration in 31% of the small reticular cells of the medulla without clinical evidence of dysfunction of respiration, and in 19% of the large reticular cells without failure of circulation. It is known that the kidney has a large margin of safety and allows for great damage before failure occurs. It is not surprising, therefore, if an organ as essential as the brain should be equally well prepared to meet contingencies. This may explain why brains can withstand periods of starvation even in utero without injury, so long as the oxygen supply, glucose, and water are maintained. With maternal infection, the fetal brain may escape either because of the blood brain barrier or because the cells survive without permanent

sequelae in most infections. Not many infections create the havoc which German measles has.

As noted earlier the neural cord is laid down early,[11 & 12] then waits for the establishment of circulation, after which there is a sudden expansion of the vestibules; the choroid plexus makes its appearance and assists with nutrition until the rest of circulation can take over. In due time, the special senses make their appearance: vision, audition, scent, touch and taste. The brain stem becomes the relay station to the cortex from all the affective inputs. The localization of these functions in the cortex is fairly well documented, so we think. However, this mysterious organ may still hold many secrets from us.

The fully developed brain has the two hemispheres separated by the Falx cerebri and these from the cerebellum by the tentorium cerebelli. The lateral surfaces of the hemispheres are designated as frontal, parietal, occipital and temporal lobes, and the insula. The medical surface has the limbic lobe. In the interior is the basal ganglia, the brain stem, and its nuclei. This is the most primitive part, but with the important functions of maintaining life and relaying the messages to the acting cortex. The surface area is greatly increased by foldings into gyri. However, the two sides of the brain are not alike, and the formation of the gyri are as distinctive as finger prints; they are not exactly the same in any two individuals.

The role of different areas of the cortex is fairly well documented, although much remains unknown. The motor cortex is located on the anterior wall of the central sulcus and adjacent portions of the precentral gyrus. The sensory area is contingent to it and posterior. In addition to these primary sensory and motor areas are sections designated as secondary sensory, premotor and supplementary motor areas. The visual cortex is located in the occipital lobe.

The cortex of the temporal lobe serves speech, reading, writing, and related functions.[13] These functions are primarily developed in the temporal lobe contralateral to handedness of the individual and usually in the left temporal but with descussation of fibers to the opposite side. A recent article by Pontius[14] attributes to the frontal lobe much of the intellectual functioning, and compares certain symptoms such as disorganization, hyperactivity and autism to the symptoms following lobotomy. The cerebellum is concerned with coordination of somatic activity, stabilization, muscle tone, and equilibrium.

The basal ganglia, putamen, globus pallidus, caudate nucleus, and amygdaloid nuclear complex is usually called the "extrapyramidal" system.[11,12] Disturbances in this area involve motor control (such as tremors, athetosis, chorea, dyskinesia, ballism, etc.)

The substantia nigra is the largest nuclear mass in the mesencephalon, extending through its length and into the caudal diencephalon. It has two distinct parts: a compact part composed of large cells with melanin granules, and a ventral reticular part. It is one of the nucleoli affected in paralysis agitans. The large cells apparently synthesize and store primary catecholamines. It is suggested that the fibers convey dopamine, norepinephrine and serotonin to terminals in the striatum.[15,16] The large white masses and bundles can be compared to cables carrying innumerable connecting fibers from the central nervous system to and from all parts of the body.

Blood is supplied to the brain by the internal carotid arteries and the vertebral arteries. The internal carotid has four branches or segments; cervical, intra-petrosal, intracavernous and supraclinoid. The intracavernous branch lies close to the wall of the cavernous sinus and the cranial nerves III, IV, VI, and part of V. The supraclinoid extends up and backward giving rise to all the major branches of the internal carotid. The vertebral arteries supply

the medulla, pons, midbrain, cerebellum and the posterior part of the diencephalon, parts of the occipital lobe and the temporal lobe. The Circle of Willis encircles the optic chiasm being formed by the anastomotic branches of the internal carotid and the vertebral arteries. The venous drainage is chiefly through the dural sinuses. These are mostly triangular in shape and have no valves. The superficial cortical veins drain largely into the superior longitudinal sinus. The deep cerebral veins drain the basal ganglia. The junction of the two internal cerebral veins form the vein of Galen. The superior longitudinal sinus is in the Falx Cerebri. The transverse sinuses conduct the blood received from the longitudinal and the straight sinuses to the internal jugular vein.

BRAIN PATHOLOGY IN NEONATAL CASES

The margin between survival and death in the newborn may be very narrow. In any study of handicapped children, therefore, it is perinent to review the pathology of stillborn and neonatal deaths, especially of those whose fetal course and development had been normal. Potters[17] extensive work in this area is reviewed and supplemented by the more recent studies of Towbin and Terplan.[18,19,20]

The hazards of delivery at term are primarily anoxia, trauma and, less often in recent years, infection. Potter describes the types of anoxia as anoxic, anemic, stagnant and histotoxic. Anoxic anoxia results from interference with the source of oxygen supply. Before birth, this may result from failure of fetal blood to be oxygenated in the placenta, or failure of oxygenated blood to pass through the umbilical cord. This may occur with premature separation of the placenta or with entanglement and obstruction of the umbilical cord. After birth, the most common cause is pulmonary pathology, usually hyaline membrane disease.

In anemic anoxia, the blood flow is adequate but the hemoglobin or red cells are insufficient to transport an adequate supply of oxygen to the tissues,

as in the hemolysis of red cells due to isoimmunization. Less frequently, it may be due to rupture of vessels in the placenta or cord. After birth, hemorrhage from any cause can produce an anoxic condition.

In stagnant anoxia, the blood does not move through the vessels adequately as can occur with maternal death, shock, or heart failure. It can occur from regional interference of circulation, with contraction of the cord, or when there is contraction of the cervix sufficient to interfere with normal flow.

Histotoxic anoxia is a disturbance in the cells of the fetus, impairing the utilization of oxygen or making it impossible. This usually results from chemical agents, or from drugs administered to the mother or taken by her.

If the lack of oxygen has been abrupt and of short duration (5 to 8 minutes is the longest for survival), the brain shows no pathology under the present methods of study. Apparently not enough time has elapsed to cause detectable changes in the cells. With longer duration of anoxia, such as may occur with premature separation of placenta, specific changes are observed. These consist of countless sharply circumscribed petechial hemorrhages, most marked in the areas of the brain adjacent to the ventricular system. With prolonged difficult labor and delivery, there is severe congestion of the leptomeninges and infiltration of red cells in the subarachnoid space and blood may be found over the hemispheres and in the sulci. It is necessary to differentiate this type of bleeding from hemorrhage due to trauma.

Although isoimmunization does not now pose the problem that it did some years ago, it should not be forgotten. Kernicterus[21] caused considerable crippling and many deaths. Potter describes the following areas of the central nervous system as those mainly involved: the basal ganglia, thalmus, dentate nucleus, flocculus, posterior vermis of the cerebellum, cranial nerve nucleoli, and the gray matter of the spinal cord. At present, apparently there

is no explanation for this selectivity. Five to ten percent of erythroblastic infants suffer damage chiefly in the form of choreoathetosis.

Death of infants due to trauma during delivery has decreased markedly in the past two decades, at least in most well-conducted maternity services. Death usually occurs from tears of the tentorium from prolonged labor, molding of the head beyond tentorial endurance, or from precipitous delivery and sudden change in pressures. When the tentorium ruptures, the rent may extend medially to the sinuses. Bleeding is subtentorial into the subdural space. If the anterior-posterior diameter is increased excessively, the vein of Galen may rupture and result in compression of the vital centers and immediate death.

A subdural hematoma usually involves the middle meningeal vessel with the blood collecting in the subarachnoid space. The pressure may destroy the underlying brain tissue and the pathology may extend into the ventricles to cause motor and sensory disturbances. Intraventricular hemorrhages usually occur in prematures. If hemorrhage occurs before labor, it may be due to a ruptured aneurysm which may produce porencephaly. Natelson and Sayers[22] report on the fate of 42 infants sustaining severe head trauma during delivery. A follow-up which included school performance was made 8 to 13 years later. Six of the 42 died within a week after admission; 5 had acute subdural hematoma, with congenital anomalies present in 3 of them, 1 was premature. Thirty six were discharged alive. Of these, 18 had fractures, 15 of which were depressed; of these children, one was severely retarded, 1 hemi-plegic-trainable, and 13 were apparently normal. Two children had linear fractures, one of whom was lost to follow-up and the other had an IQ of 84. The one patient with basal fracture had a facial weakness that cleared by the 7th day and his progress is satisfactory. Of the 24 who had hematomas, 50% of those with acute subdural hematomas are dead and only 1 had a normal IQ, 3 are retarded-trainable, and 1 is

severely retarded. Of 13 with chronic subdural hematomas, only 3 are known to have normal mentality; 2 were retarded but considered trainable.

Even though the blood-brain barrier may protect the embryo and fetus from many types of infection and injury, this protection is not complete, as the tragic story of German measles illustrates. Syphilis and gonnorhea, once thought to be eliminated or very nearly so, again threaten the newborn. Cytomegalic inclusion disease and toxoplasmosis also may cause severe pathology in the fetus, but fortunately these conditions are rare. With the advent of antibiotics, some organisms, notably the staphylococci, developed resistance and posed a new hazard in the nurseries. Sepsis due to other organisms, such as streptococcus, B. Coli and pneumococcus, occasionally occur and, even if successfully eliminated by the use of antibiotics, may leave the infant with serious sequelae. Meningitis in the newborn or young infant has caused pathology varing from microcephaly to dyslexia.

Autopsy studies of children and adults with cerebral palsy and mental retardation[23] show the end-results of the pathology described by Potter, Towbin, and others, such as petechiae and infarctions of the periventricular system, striatum, thalamus, and basal ganglia. White fibers resembling marble ramify through the basal ganglia and replace degenerated tissue. This area corresponds to the territory drained primarily by the venous system of the periventricular and dorsal regions. The great vein of Galen drains the frontal, parietal and occipital lobes. Birth trauma and infection result in pathology of the cortex, primarily the gray matter. This may consist of general, hemi- or lobar atrophy. Histologically, it may be sclerotic or show cystic degeneration.

CORRELATION OF CERTAIN CLINICAL CASES WITH ETIOLOGY AND PATHOLOGY

Can the history and symptoms of patients seen in a clinic for the handicapped be reconciled with the birth history, the brain anatomy and the pathology? As

noted earlier, this study is concerned with those children whose physical and mental development presumably were normal up to the time of injury. No attempt will be made to illustrate all the possible injuries, but to select a few that are pertinent to observations made and to questions raised thereby.

Trauma has been reviewed carefully in 42 cases by Natelson and Sayer,[22] as noted above, and their findings confirm our observations. Some of the sequelae following strangulation[2] was reviewed in the author's previous study, and they also will be omitted here. One of the intriguing questions in brain injury is how and why the intelligence of some severely handicapped individuals is normal or superior, while others with minimal symptoms suffer marked interference with intellectual performance. As examples of high intelligence with severe motor involvement, the following case histories are given:

A young lady of 22 years stopped by recently to ask if I thought she might study medicine. She has a severe athetoid quadriplegia with rather marked speech difficulty due to contortions of the facial muscles. She had graduated from college with top honors and had been elected president of her class. This girl was from a first pregnancy of a young mother who had been admitted to the hospital in normal labor. She was given spinal anaesthesia,but when turned on her back to prepare for delivery, she went into severe shock. Her blood pressure dropped, her pulse was barely perceptible and the physician feared her death. She was returned to the side position, given stimulants and resuscitation. She revived and was delivered of a baby girl weighing 8 lbs 12 ounces. The infant was quiet and limp, but when stimulated cried normally. She fed poorly in the nursery and remained rather atonic and quiet. She left the hospital with her mother and was thereafter followed at home and in the office. She had no physical stigmata. She continued to feed poorly but it was noted that at about 10 months she reacted to strangers by crying, a good sign. It was apparent that she was athetoid. She attended the nursery school for cerebral palsied children

from the age of 2 years and later a grade school for the handicapped. She did not walk until she was 6 years of age, and still has a very marked athetoid gait. The family left the community when she was about 12 years of age and she was not seen again in the office until she came to consult about her plans to study medicine. She is a delightful person, well adjusted, ambitious, obviously highly intelligent, and serene.

Another child, a boy now 11 years of age, has the same symptamology as the previous child, but more severe. He is a quadriplegic, with mixed rigidity and anthetoid movements, who cannot get about by himself and who has virtually no speech. It was also noted that he reacted with crying to strangers between 9 and 10 months of age. His mother constantly remarked that he was a very bright child. He has been tested in a number of sessions over the years by Dr. Morrison Gardner of the Child Development Center at Childrens Hospital in San Francisco, who has had many years of experience working with the handicapped and felt challenged to try to determine this child's intellectual capacity. He reorganized his tests into multiple choice questions or they were read to him and he was asked to push the block with the correct number to the question. The mother was not in the room and the child was supported by an attendant in the Center. The boy was given an IQ of 140, but Dr. Gardner thought that was a conservative estimate. This boy was the product of a twin pregnancy. At 5 months gestation, the mother had an episode of vaginal bleeding but carried the pregnancy past 7 months. The one twin was dead and, from appearances, had been for some time. The proband was described as barely viable.

My first acquaintance with the athetoid phenomenon was in college when, from my sorority window, I could see a severely athetoid young man struggle his way to classes. Later, I learned that he was Earl Carlson,[24] author of t he book, "Born That Way." He also was responsible for stimulating national concern

for the cerebral palsied patient.

In Ireland, some years ago, I met Dr. Robert Collis, who showed me another book written by a cerebral palsied patient who was so handicapped that the story of his life was told in a book entitled, "My Left Foot". That was how he actually penned the story.

When reading the literature on the development of the brain, I was impressed by the proliferation of the vessels around the site of injury and also around areas of destruction. The question arose, "Is there a very narrow margin in hypoxia, where there might actually be a stimulation of the uninjured parts of the brain to perform better than average?" This to me, appears a distinct possibility and should be investigated further.

Another problem is the so-called "Minimal Brain Damage Syndrome", also known by many other names. The following history illustrates such a case. Although miningitis primarily involves the meninges and prompt treatment will usually mean recovery without sequelae, the infant and young child may not be that fortunate. An infant with pneumococcus meningitis and resulting microcephaly was seen and reported by the author in 1949.[2] In another infant, influenza meningitis resulted in recovery and the "Minimal Brain Damage" syndrome. This young man, now 21 years of age, has a true dyslexia . He has slight difficulty with writing and with speech but he has been fortunate in his training in a small town in Minnesota. His first years in a regular school were difficult because of his inability to learn to read. However because he loved sports, he was made custodian of the equipment and thus participated in all the games and won his athletic letter. At the high school level, a retired Army officer initiated a rehabilitation program for children with learning problems and this boy was admitted to the classes. Mornings were spent in the class room, but not in

the usual manner. The time was devoted to acquiring, by any means possible, enough skills to function on the outside. This meant a minimal vocabulary and recognition of a few essential words and numbers. Also considerable time was spent in learning the amenities, i.e., how to meet and greet people, how to introduce yourself or others, courtesies, grooming, etc. Afternoons were spent in medicine what we might call clerkships. Through arrangements with businesses, shops, banks, mills, stores, hospitals, senior citizin homes and even the morgue, the students were rotated for certain periods of time through these agencies and were taught skills. They were taught principles of banking, making out checks, depositing them and saving; first aid and nursing principles in the hospital and nursing homes; instructed in waiting on tables, taking orders etc., in the cafeterias; and in the morgue they encountered death and how to deal with it. The philosophy behind the program was an emphasis on the positive learning in all areas and a de-emphasis on the negative side. The method had a very positive effect on the students. It avoided years of stress in situations where they could not compete in the class room. In all other activities, they participated with the rest of the high school students. The young man cited is now holding down a good job.

Poor nutrition[25,26] as a factor in brain damage has been given a great deal of attention the last decade. A review of starvation was made in the previous paper with the conclusion that it is not a factor in congenital anomalies although it plays a role in late pregnancy and especially in certain specific deficiencies, such as vitamins. Its role in the growth of the brain is still controversial. To illustrate this occurence, an opportunity presented itself in 1953 when a baby girl was born at term with marked intrauterine growth retardation. This infant weighed 3lbs 4 ounces at birth. The placenta was markedly infarcted and the infant was emaciated to an extreme degree and described

by the intern as resembling a little old woman. There were no physical anomalies and none of the stigmata often associated with congenital defects. She did not react as a premature, she took her bottle eagerly and gained to 5 lbs in 3 weeks. She was followed to the age of 5 years and appeared normal in all respects, although on the low side of growth curve. While those who work with handicapped children are the first to point out that it is necessary to wait for school age to be sure about learning difficulties, pediatricians recognize normal behavior and other parameters of growth and this child fell well within the normal. Also these workers are well aware that the IQ might have been higher under more favorable circumstances.

Although I am very reluctant to extrapolate from animals, an article on the lungfish is interesting.[27] When deprived of water in its natural habitat it burrows head first into the soft mud to a depth of 18 inches or more. It settles down with its head pointed upward, its body loops in the middle and doubles back, so that the tail covers and protects the eyes. Its muscle tissue is the source of its food. Lungfish have been known to lose ½ of their weight and survive. They have been kept also in blocks of hardened mud for four years and come out"thin, emaciated, hungry, but perfectly healthy". Our little girl had probably lost half her weight.

In contrast to the above tales, some equally fabulous escapes from injury under dramatic circumstances can be cited. When working in the communicable disease division of a children's hospital, a pregnant woman near term was admitted with pneumococcus meningitis and died within hours. The obstetrical department was alerted, performed a caesarean after her death and delivered a normal infant. The child was followed for only the first year but appeared to be normal in all respects.

Another mother 27 years of age was diagnosed November 1952 as lymphocytic

leukemia[28], with a hemoglobin of 7.8 gm and rbc of 2,100,000. She was 3½ months pregnant and dated her poor health to July 1952 (before her onset of prenancy). During the next several months, she was treated with transfusion, chemotherapy, and cortisone in an attempt to control her leukemia. On March 18, 1953 a caesarean section was performed but the mother died 9 days later. The infant cried immediately after delivery and his blood and marrow were normal. This child was followed for 4 years and his development and performance were normal.

When poliomyelitis was prevalent, a number of pregnant women were admitted and Dr. Schaefer and Shaw[29] who reviewed the cases and found 18 pregnant women among 500 admissions. Seven of these were in the first trimester, 3 went to term and delivered normal infants; 3 aborted and 1 delivered a premature infant that died (she had placenta previa). Of 7 who developed polio during the second trimester, 5 had full term normal babies, 1 delivered twins at 8 months, she had extensive paralysis and bulbar symptoms which required the respirator, where she spent half her pregnancy. Her twins were normal, even though they were exposed to the virus of polio, the possibility of low oxygen in a respirator, and the lack of maternal exercise for half the pregnancy.

REFLECTIONS

These two studies have been undertaken in an attempt to provide a partial answer to the statement, "We don't know the cause." There is a great deal we do not know, but this is no reason for putting our heads in the sand and pleading ignorance to all causes. We know a great deal about embryology, genetics, and organogenesis and we also have considerable data on interferences with the normal course of growth and development. A review of these factors seemed timely.

So many defects are associated with mental retardation. The difference between these children and those who had developed normally in gestation and then been injured was apparent early in our studies at the Child Development Center. It seemed pertinent to call special attention, therefore, to the possibility of a high intelligence to survive in a child who has suffered severe and totally incapcitating physical injury at birth. It is easy for both educators and physicians to overlook this possibility when individuals can not speak up for themselves. The plight of such individuals is great and deserves great sympathy and understanding from workers in the field. We need to devise a life style that will help them to reach their best potential.

From the literature, children with learning problems are getting a great deal of attention but, with our present educational hangups, other training has been sorely neglected. It is time that we analyze these children for their positive assets and train them without destroying their morale and self-image by pursuing the three Rs through the grades beyond, but without otherwise accomplishing any worthwhile advancement for them.

With the present emphasis on nutrition and growth, including that of the

brain and without appreciation of the uniqueness of this organ in structure as well as function, there is a danger that another group of children will be assumed to be retarded because they have suffered starvation through some period of their development. Let's recall the lungfish.

I would like to see a large model of the brain and its communicating system set up in our medical school corridor. Even though our ignorance is still colossal, if we passed it every day and thought about it, we might get some inspirations. Hopefully, we are still evolving.

REFERENCES:

1. Thelander, H.E.: Interference with organogenesis and fetal development, Texas Rept. Biol. Med., Vol. 31, No. 4, 1973.

2. Thelander, H.E., and G. Goebel: Microcephalia as a complication of pneumococcic memingitis, Amer. J. Dis. Child. 77:642-646, 1949

3. Snell, R.S.: Clinical Embryology for Medical Students, Little Brown Boston, pp. 11-17, 60-61, 1972.

4. Ibid. pp. 60-61.

5. Mayo Clinic, Rochester: Clinical Examination in Neurology, W.B. Saunders Phila., 3rd ed., 1971.

6. Klosovoskii, B.N.: Institute of Pediatrics of Academy of Science, USSR, Lomonodorski, Moscow, vol. 261.

7. Klosovoskii, B.N.: Blood Circulation in the Brain, A.D. Behmoaram (transl.) Israel Program for Scientific Translations in Jerusalem, 1963, distributed by U.S. Dept. Commerce, Washington, D.C., 1963.

8. Ochs, S.: Elements of Neurophysiology, JohnoWiley & Sons, Inc., N.Y., 1965

9. Minkowski, A., J.C. Laroche, J. Vignaud, C. Dreyfus, S.S. Brisac, A.Dargasses: Human Development, F. Falkner, ed., Development of the Nervous System in Early Life, 1966, S. Karger, Basel.

10. Baker, A.B., H.A.Matske, J.R.Brown: Poliomyelitis: Bulbar poliomyelitis, a study of medullary function, Arch. Neurol. Psychol. 63:251-281,1950.

11. Carpenter, M.B.: Core Text of Neuroanatomy, Williams & Wilkins, Baltimore, 1972.

12. Peele,T.L.: The Neuroanatomic Basis for Clinical Neurology, McGraw Hill, N.Y., 1961.

13. Penfield, W., R. Lamar: Speech and Brain Mechanisms, Princeton Univ. Press Princeton, N.J., 1959.

14. Pontius, A.A.L: Dysfunction patterns analagous to frontal lobe system and caudate nucleus syndrome in some groups of minimal brain dysfunction, J.Amer. Med. Wom. A. 26:285-292, 1973

15. Albers, R.W., G.J.Siegel, R.Katzman, B.W.Agranoff (eds.) : Basic Neurochemistry, Boston, Little Brown, 1972.

16. Chusid, J.G.: Correlative Neuroanatomy and Functional Neurology, Lange Palo Alto, 1970.

17. Potter, E.L.: Pathology of the Fetus and Newborn, Yearbook Publishers, Chicago, 1952.

18. Towbin,A.: Mental retardation due to germinal matrix infarction, Science 164:156-161, 1969.

19. Towbin,A. : Central nervous system damage in the human fetus and newborn, Amer. J. Dis. Child. 119:529-542, 1970

20. Terplan, K.L.: Histopathological brain changes in 1152 cases of the perinatal and early infancy period, Biol. Neonat. 11:348-366, 1967.

21. Sass-Kortsak, A. (ed.): Kernicterus, Univ. Toronto Press, Toronto, 1959.

22. Natelson, S.E. And M.P.Sayers: The fate of children sustaining severe head trauma during birth, Pediatrics, 51:169-174, 1973

23. Malamud, N. : Atlas of Neuropathy, Univ. California Press, Berkeley, 1957.

24. Carlson, E. : Born That Way, John Day, N.Y. 1968.

25. Moncha, S.L.: Malnutrition and Human Development, C.C. Thomas, N.Y., 1972.

26. Winick, M.: Nutrition and Development, New York, Wiley, 1972.

27. Coates, C.W.: Slowly the lungfish gives up its secrets, N.Y., ZOological Society 40:25-34, 1937.

28. Bierman, H, P.Aggeler, H.E. Thelander, K.H. Kelly, F.Cordes: Leukemia and pregnancy, JAMA 161:220-222, 1956.

29. Schaeffer, J., E:B. Shaw: Poliomyelitis in pregnancy, Calif. Med. 70:16-18,1949.

Causes of Birth Defects

Letter to the Editor. When working with brain-injured children, the dichotomy of severe mental retardation versus genius among them was a constant enigma. In my extensive review of the literature[1] after retirement, I came across the following statement by Klosovoski[2] in his study of blood circulation of the brain , " The metabolic products of these cells and the waste products resulting from their disintegration (mainly their nuclei) are powerful stimulants which trigger the vigorous and massive new growth of vessels from the pial membrane into the wall of the primary cerebral vessels as observed by us during the second half of pregnancy." Is it possible that this invasion of blood vessels into an injured area may be a stimulant to adjacent tissues still in the developing stage?

I will cite two instances as examples of severely injured children followed from birth with marked physical impairment but exceptional minds. CW now twenty two years of age, a girl who graduated at the head of her clas with honors and who wants to study medicine, is a severe athetoid quadriplegia with marked interference with speech. She was the first born of a young mother admitted to the hospital in labor and givena spinal anaesthia. When turned on her back for delivery, she went into severe shock, her blood pressure dropped, her pulse was barely perceptible and the physicians feared she would die. She was turned to her side, given stimulants and resuscitation. She revived and delivered a baby girl weighing 8 Lbs. 12 Oz. The infant was quiet and limp but cried when stimulated. She remained quiet in the nursery and fed poorly, however she went home with the mother, and there after was allowed in the home and office. She had no physical stigmata. At ten months she was responsive and began to react by crying to strangers, a favorable developmental sign. She was then also recognized as being athetoid. She attended the Cerebral Palsy nursery school until old enough to be admitted to a public school for the handicapped. She did not walk until 6 years of age and as noted above is still very handicapped.

The second case, NC, a boy now twelve years of age, has similar symptomatology but more severe. He is a quadriplegic, with mixed rigidity and athetosis and no speech. He also reacted with crying in the presence of strangers between nine and ten months of age. His mother maintained that he was a very bright child. He was the product of a twin pregnancy. At 5 months gestation, the mother had an episode of bleeding but carried the pregnancy past seven months. One twin was dead and from appearances, had been so for some time. The proband was described as barely viable. Dr. Morrison Gardner of the Child Development Center at Childrens Hospital, had had many years of experience working with the handicapped felt challanged to try to determine this childs intellectual capacity. The testing was done during many encounters and with ingenious techniques. The boy was given IQ of 140 but Dr. Gardner stated that he thought this was a conservative estimate.

The major factor in both these cases was undoubtedly hypoxia. Anoxia of more than 5 to 8 minutes is fatal. There is, therefore, a very narrow margin between survival and death. Dr. Potter [3] describes the pathology of fatal cases of anoxia as, " Sharply circumscribed petechial hemmorages most marked in the area of the brain adjacent to the ventricular system". This pathology corresponds with the symptoms of athetosis.

In discussing these findings with colleagues, i frequently get a comment suggesting that maybe he or she was brighter and 10 or 20 points lowering of intelligence would still leave a child normal or maybe above. This seems very unlikely as an explanation. I would like some comments from physicians with experience in the field,- neuropathologists, embryologists.

H.E. Thelander M.D.
Child Development Center, Emerits
Childrens Hospital, San Francisco

12/ 28/ 74.

References

1. Potter, E.L. Pathology of the Fetus and Newborn. Year Book Publishers.

2. Thelander, Hulda E. Interference with Organogenesis and Fetal Development. Texas Reports on Biology and Medicine, Vol. 31, No. 4. Winter 1973. pp 791- 861.

3. Klosovoskii, B.N. Blood Circulation in the Brain/ Translated from the Russian. Published for he National Library of Medicine, U.S. Public Health Service by the Israeli Program for Scientific Translations. 1963. pp 267.

Gentle Hand With The Handicapped

By CLAIRE LEEDS

GIVING BIRTH to a handicapped child is something people say can't happen to THEM—until one day it does!

If you should be one of these parents, what would you do? Where would you turn for help?

One of the most hopeful answers ever offered here is the new March of Dimes Birth Defects Special Treatment Center at Children's Hospital. There a dedicated staff of specialists is prepared to offer parents and their children all humanly possible help and encouragement.

Inspiration behind this remarkable facility is Dr. Hulda E. Thelander, its director, a noted neurological specialist and pediatrician.

Long known as a source of strength to parents of handicapped children, Doctor Thelander now is able to reach many more, thanks to March of Dimes financing.

DOCTOR THELANDER is a slight, soft spoken woman with a fierce determination to help handicapped children get an even break.

The big difference the March of Dimes treatment center is making toward this goal is in offering parents one place to take their "thousand questions about their handicapped child."

The child born blind, or with cerebral palsy or other congenital defects, usually starts out in life not with one, but a whole series of problems. In the past, his parents have been forced to shop around from doctor to doctor and clinic to clinic in search of help.

Now they may take him to the March of Dimes Center where, in a setting much like a cheerful nursery school he gets whatever he needs, from thorough diagnosis and medical help to therapy and psychological testing.

FOR HIS worried parents, there is the wise and compassionate counsel of Doctor Thelander and her staff of doctors, therapists, psychologists and social workers.

What do you tell the parents of a child with a congenital handicap?

"You try to help them understand that this is first a child, then one with a handicap," explains Doctor Thelander. "That, like any other baby, he needs love, attention, and the joy of normal experiences."

THE CHILD born handicapped is not conscious that he is "different" because this is all he has ever known. Thus it is essential to see his normal side and help him

(Continued on Page 22, Col. 8)

DR. HULDA E. THELANDER
"Look at handicapped child's normal side"

Handicapped

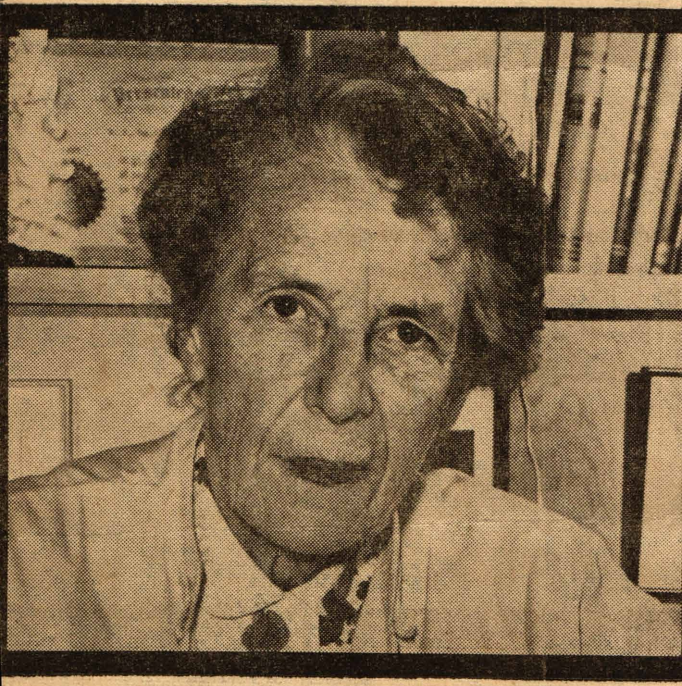
(Continued from Page 21)

to face the future with confidence, Doctor Thelander believes.

The parent must try to find a realistic goal for the child, to show the child that while there are certain things he cannot do, there are others he CAN do.

Doctor Thelander recommends a judicious use of examples of people who have overcome handicaps with success—like Franklin Roosevelt. But she warns against overdoing it, such as "insisting that every blind child can become another Helen Keller!"

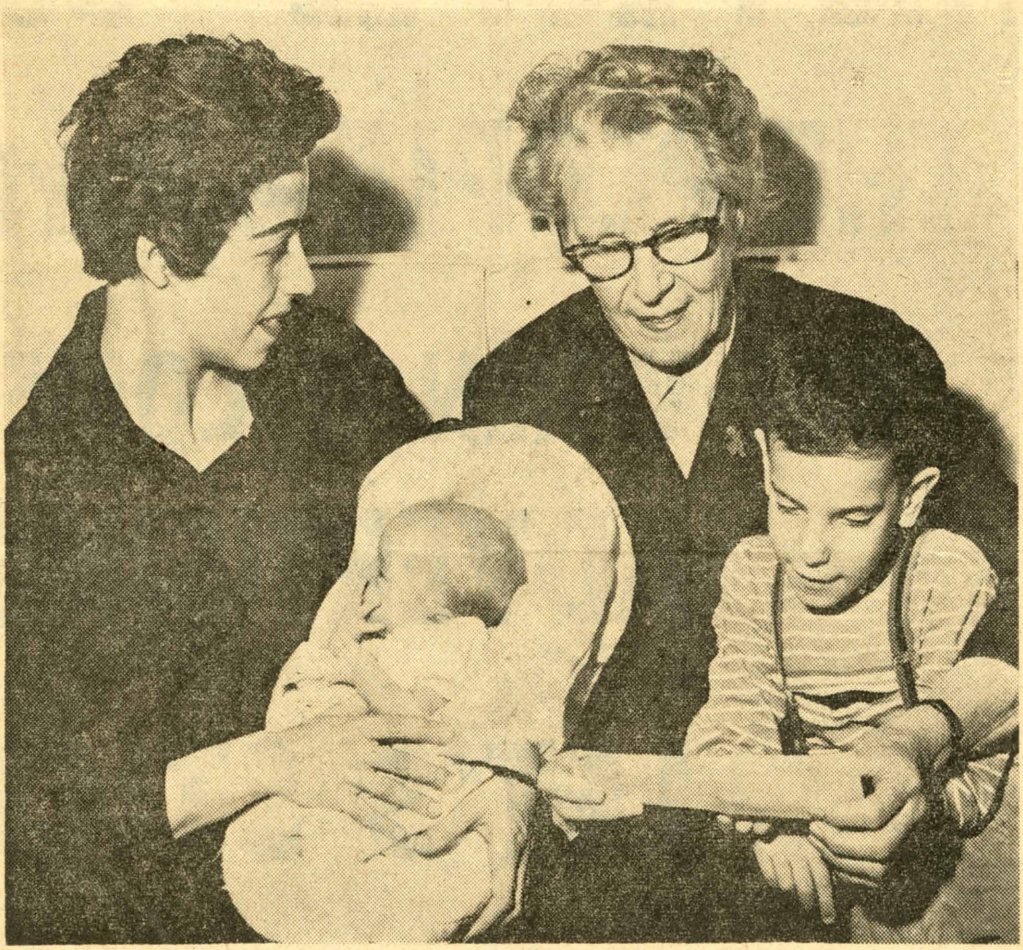
Love her
nice nice nice nice nice



DR. HULDA THELANDER

pediatrician and inspiration to parents of handicapped children

GHT EDITION



BIRTH DEFECTS SPECIALIST ON TV

Dr. H. E. Thelander, director of the March of Dimes-supported Birth Defects Special Treatment Center at Children's Hospital, is shown here (right) with two young patients at the center, and a young mother. Mrs. Jack Jones holds baby Joyce, while David Palmer, 6, looks at a March of Dimes check. Dr. Thelander will be the guest scientist next Monday night (Jan. 15) on "Science in Action," KRON at 7.

The March of Dimes-supported Birth Defects Center is the first in California and the sixth in the United States. It was officially opened last November. Dr. Thelander has devoted a good part of her life to working with children born handicapped, and with their parents.

"Working with the parents is just as important as working with the children, perhaps even more important," said Dr. Thelander. "A child with a built-in defect has never known what it is to be normal. The child gets his image of himself from the reactions of others.

"Therefore, it is important that the adults around the child react in such a way as to give him as near a normal image of himself as possible. A child grows up thinking he is what the world around him thinks he is. Making surroundings as nearly normal as possible is the first step to success."

Undecided About

Ikebana International

Hope for Cancer Cure Told

"I MAY BE too much of an optimist but I believe that someday we will have drugs that at least will cure some cancers," Dr. Hulda E. Thelander told 150 delegates to the forty-ninth annual convention of Phi Beta Psi Sorority Monday.

Doctor Thelander, chief of the department of pediatrics at Children's Hospital and an associate clinical professor at UC, was the speaker at the opening session of the national convention at the Mark Hopkins. Her topic, "Cancer in Childhood," was of special interest to the sorority, which made cancer research its national service project in 1942.

The sorority, which numbers 2,900 members in ninety-five chapters, sponsors two cancer research projects at the University of Pennsylvania. Seven of the chapters are in California.

Attending the convention was one of the three founders, Miss Edith Wolf of Huntington Park. National president is Mrs. C. R. Vancil of Springfield, Illinois.

The convention closed last night with a 7 p. m. banquet and candlelight ceremony followed by a dance in the Mark Hopkins' Peacock Court.

Smart Set
FEATURES
San Francisco Examiner 18
Thursday, July 14, 1955 CCCC*

HANDICAPPED CHILDREN IN SAN FRANCISCO

The total number, as listed by the San Francisco Survey of 1954 and 1955, is 21,000. The majority of these were noted as minor but long term handicaps not incapacitating the child. The rest of the figures below were taken from the report of 1955 plus reports by the Recreation for the Handicapped Society in 1959 and from Dr. Babow's report on Rehabilitation in 1960. The numbers are therefore in round figures because each group used a different age. Thus the 1955 survey divided them at 15 years, the Recreation Club divides them at 18 years, and another report divided them at under 25 years. The figures are estimates but they should be fairly accurate for about 21 years or under. It would seem to me that 21 years would be a good dividing point as this is the age up to which Crippled Children Services take them and it is the age at which the Academy of Pediatrics divides them.

Speech --- ---	6,000 plus
Mental Retardation --- ---	2,000 to 3,000; of these 200-300 are severe
Hearing Impairment	500 plus -
Orthopedic Impairment	500 to 600 -
Cardiac, including rheumatic	300 plus or minus -
Visual Handicaps (the severer type)	150 to 200 -

Others include multiple handicapped children, anomalies, chronic asthma, cystic fibrosis, etc., etc.

There are in San Francisco 15,000 children under ANC. These might be listed as the deprived group.

January 1961

1963

A PROPOSAL FOR AN EXPERIMENTAL SCHOOL TO STUDY LEARNING PROBLEMS IN CHILDREN

In the past ten years the medical and ancillary staffs at the Child Development Center at Childrens Hospital have studied approximately 1000 children with either injury to or developmental anomalies of the central nervous sytem. The study includes a few children with apparent learning problems who turned out to be children with superior intelligence who were merely bored.

This study has been made against a background of thorough familiarity with the normal child.

There is a great need in San Francisco for a school facility where educators can cooperate with the staff of the Child Development Center in exploring in depth the learning process with the aim of developing more effective teaching techniques to help children with learning problems.

NORMAL VARIATIONS:

1. Children vary in the rate at which they learn new things.
2. They vary in the age at which they are ready to learn new subjects.
3. They vary in the manner in which they acquire new knowledge.
4. They vary in ability to comprehend new subjects.
5. They vary in curiosity, motivation, persistence and stamina.

ABNORMALITIES IN LEARNING:

Children who are born with an anomaly of the brain or who have acquired an insult to the brain due to disease or injury have all the variations of normal children. In addition they almost always have specific learning disabilities. We accept that elderly people who have a stroke may lose the ability to speak, that they may have erratic memories, that they may be able to understand but cannot choose the right word or the right name, etc. Children with abnormalities of the nervous system may have all the difficulties that an adult with a stroke has and many more. Furthermore, they are without the experience and background that an adult who sustains an injury has to build upon. THIS IS THE FIELD THAT NEEDS TO BE EXPLORED. THERE IS NO QUESTION THAT MANY CHILDREN ARE CONSIDERED MENTALLY RETARDED WHO HAVE SPECIFIC LEARNING DISABILITIES.

TYPES OF DISABILITIES:

1. Inability to speak (aphasia)
2. Inability to learn to read (dyslexia)
3. Inability to learn numbers (dyscalculi)
4. Inability to learn to write (often a visual perceptual problem)
5. Blind children who do not use touch to help themselves
6. Hyperkinesis. Children who are overactive, have a short attention span, erratic memory, compulsive behavior.
7. Children with total retardation (mongols).

THE CONSEQUENCES:

Many of these children have normal or even superior ability in some fields. It is now recognized by pediatricians, neurologists, psychologists and social workers in this field that some of these children as a result of the failure of adults to recognize the problem, become truants from school and eventually delinquents. Others become emotionally disturbed or even psychotic, still others make a relative adjustment but do not function up to their potential.

n

THE PROPOSAL:

A Private school preferably near the Child Development Center that would work with the staff of the Center in studying and exploring means by which these children could be helped. Some methods already suggested are:

1. Learning machines.
2. Programs re-structured along the methods of the material used in learning machines.
3. Re-inforcement of learning by the use of a combination of methods (such as visual, auditory, and tactile).
4. Application of known techniques to new situations, such as trying Braille for children with dyslexia.
5. Strengthening good areas to the maximum and see if this improves the deficient areas by overflow as it were.

PERSONNEL:

Such a school calls for teachers with creativity, imagination and optimism as well as faith in the children. The teachers should have freedom to try new methods. It would be very interesting to have a class of superior children in the school in order to study their learning methods and also to use them at times to teach or possibly to help adults understand the children better.

H. E. Thelander, M.D.

February 11, 1963

A Preschool Cerebral Palsy Program

H. E. Thelander, M.D.

THE HABILITATION AND REHABILITATION of the brain-damaged infant is a challenge to modern medical disciplines.

The program at Children's Hospital, supported by a grant from the San Francisco United Cerebral Palsy Association, was started in August 1952. Since then many requests for information have been received and a presentation of our experience in the organization of such a program seems timely in order that others contemplating work in this field may profit from what we have learned. For the sake of clarity, the project will be presented under the following divisions: Preparation, Directorship, Organization, Education, and Research.

Preparation

The most important single factor in the preparation for work with the brain-damaged infant and child is a thorough understanding of normal growth and development, physically, mentally, and emotionally. The staff at Children's Hospital was well prepared in this area through group discussions on childhood ecology which were held weekly over a period of several years. In these sessions, not only have all aspects of normal growth and development been discussed, but also the education of handicapped children and community resources for their care have been thoroughly explored.

Habilitation and rehabilitation of the brain-damaged infant and child calls for teamwork, and that team must be larger and more diversified than for almost any other medical problem. Therefore, it is especially important that discussions at the staff level be conducted with honesty and that all essential differences of opinion be resolved before the program is launched. Our seminar had also prepared us for this aspect of the work.

Dr. Thelander is Chairman of the Department of Pediatrics, and Director of the Preschool Cerebral Palsy Program, Children's Hospital, San Francisco, California.

Directorship

The chief of the department of pediatrics is medical director of the program on a volunteer basis. The executive director, who is by profession a psychologist, is on a full time basis.

Organization

The type of local organization should be determined in part by the local conditions. In San Francisco, a program for the school-aged child had been conducted for several years under the auspices of the Division for Crippled Children of the State Department of Public Health and the University of California Medical Center. It was possible, therefore, to limit the program at Children's Hospital to the young child, primarily under 3½ years of age. Furthermore, it was desirable to set up a program that would appeal to doctors in practice and to pediatric clinics in the city as offering a service not given elsewhere. At the same time, the services should not interfere with the doctor-patient relationship, or replace medical clinic services in other hospitals. This feat was accomplished by borrowing from the pattern and experience of tumor boards. The program is as follows:

1. A clinic for the admission, evaluation, and follow-up of patients.
2. A therapy program for
   a. infants and others not ready for nursery school.
   b. nursery school children.
3. Consulation services
   a. at specialty clinics.
   b. at a monthly neurologic conference.
4. Group work
   a. with mothers of the nursery school children.
   b. in staff conferences twice weekly.
   c. in parent-faculty organization.

The ancillary staff consists of a physiotherapist, an occupational therapist, a speech therapist, a nursery school educator, a social worker, and a psychologist.

The medical staff has representation from the following departments: pediatrics, psychiatry, physical medicine, orthopedics, neurology, neurosurgery, E.N.T., eye, and speech.

The Board of Directors of the hospital has appointed a committee that has given invaluable aid and support to the program, acting as liaison between the hospital and the United Cerebral Palsy Association. A committee on nursery schools of the UCPA has assisted in the development of the nursery school therapeutic program. Although the financing of the program is in the form of a grant to Children's Hospital from the local UCPA, a careful, detailed budget based on actual costs has been worked out by the hospital administrator and the committee of the board of directors.

1. The Admission Clinic

New appointments made through the office of the executive director; patients are seen only on referral from a doctor or a medical clinic. When inquiries come direct, or through nonmedical persons or agencies, the director clarifies the methods of referral. First appointments are to the admitting clinic. Registration is completed and a social history is taken on the first visit; if not already on hand from the referring physician or clinic, a summary of the previous medical record is requested. The examining pediatrician evaluates the patient and completes the record by examination, measurements, and the assemblage of information already available. The patient is then routed into the proper therapy channel.

2. The Therapy Program

a. Children under 18 months of age and others not suitable for the nursery school are seen in physiotherapy and occupational therapy for treatment; instruction is given to mothers in techniques that can be carried out in the home. As the patient advances, return visits are made as often as necessary for evaluation and for teaching new exercises and techniques. The occupational therapist makes home visits to supplement the training. A great effort is made to enlist the parents in the therapy program, both by home visits of the occupational therapist and through parent-faculty meetings.

b. Registration in the therapeutic nursery school. At present, the nursery school has three sessions a week of 2½ hours each. Some children attend only once weekly, others twice. Lack of space has limited more frequent sessions. A nursery school educator is in charge. A scheduled program including physiotherapy, occupational therapy, and speech therapy is conducted with the children. The greatest functions, however, of the nursery school are the socialization and the opportunity afforded the handicapped child to develop autonomy within the limitations of his handicap. The mothers are permitted to be present or are within call until such a time as the children are happy without their presence. The children gradually accept the absence of the mother, relax, play, and attempt to imitate and perform what other children in the group are doing.

3. Consulation Services

a. The Specialty Clinics. Since these patients are referred by physicians or clinics, any special work that becomes necessary is discussed with the referring person or agency; management is thereby determined, whether this be orthopedic care, neurosurgical consultation, eye examination, or hearing tests. However, in order to carry out the therapy program, both at the clinic and in the nursery school, the advice of a specialist is frequently necessary. For this purpose, the consultant in physical medicine attends one clinic session a week, the orthopedist attends four sessions a month, and the neurologist is present three times a month. There is available to the clinic a physician trained in speech and hearing.

b. The Neurologic Conference. The neurologic conference, held once a month, is conducted much like a tumor board. Problem cases or those of especial interest are presented to the group by the referring doctor whenever this is possible. In attendance at the conference are representatives from the specialties of pediatrics, orthopedics, neurology, neurosurgery, physical medicine, E.N.T., eye, speech, psychiatry, and all the ancillary services. House staff, students, and guests are welcome to the meetings. Suggestions and recommendations are made. Often a group of patients with similar handicaps, such as hemiplegia or ataxia, are discussed, but usually even then there is individualization and specific suggestions and recommendations are made for each child. This has been a very valuable teaching conference and is well attended by house and visiting staff.

4. Group Work

a. Mothers of the nursery school children. During the nursery school period, the mothers meet for one hour as a group. The psychiatrist, psychologist, and the speech therapist are usually present at these sessions. If the parent desires a personal interview with the psychiatrist, it is then made by special appointment.

b. Staff Conferences. The professional staff, ancillary and medical, meet at the end of the nursery

school session for a group discussion of general problems as well as special problems related to individual children in order that a concerted effort can be carried out for the benefit of the program and each child.

c. Parent-Faculty Organization. With a little guidance, the parents organized their own group and usually suggested the subjects to be discussed at their meetings. These meetings are held once a month, business is transacted, light refreshments are served, and a program is presented. The parents have appointed one subcommittee to plan the programs, another to contact and welcome new members; some of the fathers have interested themselves in making articles for the nursery school, including a question box for parents. To date, the programs have been mostly educational. There has been a request to present Gesell's film on "Normal Growth and Development." A film on "A Place in the Sun," which describes a school for spastic children, has also been given. There have been talks by the neurologist, the neurosurgeon, and other staff members in the specialty fields.

Education

Participation in an educational program was thrust upon us almost before we were ready to receive the students, thus indicating the great need and demand for educational opportunities in this field. During the summer session at the local teachers' college, 55 students divided into groups of 8 or 10 visited the nursery school for half day sessions. Student nurses, teachers, speech therapists, and interns in occupational therapy and physiotherapy have participated in the program. There have been many other visitors.

Research

In the field of research, we have been handicapped and delayed by lack of funds, equipment, time, and personnel. There are many areas outlined which are yet to be explored. Some members of the staff are conducting research on the brain-damaged child which will eventually accrue to the benefit of our children. Some of the patients are included in a nutritional study now under way in the department of pediatrics. Preliminary steps have been taken to explore the problem of respiratory difficulty in severe brain-damaged cases.

The facilities at Children's Hospital, which include a contagious disease department, are particularly valuable for the detailed pathologic study of fatal cases of acute nervous system diseases, such as poliomyelitis and encephalitis. Such studies need to be continued in order to increase our knowledge of the localization and function of the different areas in the nervous system.

Discussion

The greatest service an established program can render to a contemplated program is to reveal the pitfalls and problems which are encountered. In cerebral palsy work, no matter how carefully forewarned about the slow progress of therapy, overenthusiasm among the parents is commonly encountered. It is well for the staff to be aware of this, to detect it, and help parents through the period of depression which follows when progress is noted to be slow. An organized parent group is a great help.

The psychology of parents, not only of the children with cerebral palsy but of other handicapped children as well, has not been sufficiently evaluated. There is a reluctance on their part to admit the existence of an emotional disturbance within themselves, and a resistance exists, probably on a guilt basis, to ask for help. Group discussion and the "peripheral" approach may succeed when direct referrals to the psychiatrist fail.

There are problems related to the patients themselves. In this young group, a variety of conditions is brought to us and the task of helping parents to find the proper solution is often difficult. There are children with cerebral palsy who, in addition, have any one or more of the following conditions: blindness, deafness, mental retardation, and psychoses. These problems have called for conferences with the personnel of many other agencies. During the coming year, we hope to open our seminar on childhood ecology to workers in other fields in order to clarify our thinking at the staff level on some of the issues that arise.

The question of follow-up beyond the age period of the children seen here has come up for discussion. As noted earlier, San Francisco is fortunate in having a resident school for children with cerebral palsy over the age of 3½ years and also a public day school for handicapped children over 3½ years.

The handicapped child with neurotic and psychotic disturbances is an especially difficult problem, and the facilities for evaluation and therapy for these are indeed inadequate.

There is need for a great deal of research in all areas of the cerebral palsied and brain-damaged child. Therefore it is recommended that at the very onset of any work with these children, an attempt be made to have observations and data carefully and accurately recorded in such a manner that they can be utilized later for evaluation and study.

Summary

An attempt has been made to summarize the organization of a preschool cerebral palsy program in one institution in such a manner that others who wish to pursue this work may find help in the positive area and may also avoid some of the pitfalls and errors to which such an organization is subject. It is indeed a challenging field and offers one of the greatest opportunities in medicine for the multiple discipline approach to the solution of the problem of the severely handicapped child.

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PEDIATRIC EVALUATION OF BRAIN-DAMAGED CHILDREN

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PEDIATRIC EVALUATION OF BRAIN-DAMAGED CHILDREN

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IN August, 1952, the prenursery school program for children with cerebral palsy was started at Children's Hospital in San Francisco, sponsored by the local United Cerebral Palsy Association.[1]

For several years members of the hospital staff, medical and ancillary, have participated in a weekly seminar on childhood ecology.[2, 3] This experience furnished the staff with excellent background and preparation for work with the handicapped child. A grant from the national U.C.P.A. in 1954 has made it possible to expand the educational program with special emphasis on the growth and development of normal children. The present report was further stimulated by a concomitant study of nutrition being conducted by Dr. Elizabeth Macaulay under a grant from General Foods.

MATERIAL

Since the opening of the cerebral palsy program August 1, 1952, 131 children have registered. Table I lists the main diagnosis for each child and Table II the secondary diagnoses. It will be noted that the average per patient is 2.3 major handicaps. The greatest number of diagnoses on a single patient was six, consisting of spastic quadriplegia, congenital hip, seizures, strabismus, mental retardation, and microcephaly. In Table III are listed the etiological factors as determined by history and physical findings on one hundred of the cases in which the study was complete enough to make an evaluation.

Several children were seen merely once in consultation and since detailed studies and follow-ups were not always available on these, analysis of various aspects may be based on different figures. This will be indicated in each instance. Of the total number of children registered, seventy-four were males and fifty-seven females. The ages varied from birth to 10 years, with 77 per cent having birth dates in 1950 or later.

Under "delayed motor development" in Table I are included infants and young children who still show only weakness of muscles, such as failure to support head or to sit up. Also included in this group are brain-injured children with mental retardation and delayed muscle development who came to the cerebral palsy clinic primarily because of failure to walk at the usual age. Apart from the above group were a few children of normal intelligence who were merely late walkers, accomplishing it at nearer 2 years than 1. Cases of central nervous system damage without motor involvement included children with speech difficulties, nystagmoid eye movements, and some with autistic or psychopathic symptoms.

The number with seizures, namely, seventeen, appears low. A partial explanation for this figure is that the children are young. Convulsions, recognizable as such, may not have appeared at this early date. In retrospect it was also apparent that the history of an occasional febrile con-

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vulsion may have been missed. The seventeen cases therefore represent children requiring treatment for seizures.

cases where the bone age was taken, ten showed rather marked retardation. This was unrelated to the severity of the physical handicap and, therefore,

TABLE I. PRIMARY DIAGNOSIS

Cerebral palsy—spastic		52
Quadriplegia	24	
Paraplegia	11	
Hemiplegia L	8	
R	7	
Triplegia	2	
Athetoid		11
Ataxic		12
Atonic (flaccid)		11
Tremor		2
Incoordination, motor		5
Delayed or retarded motor development of neurogenic origin		24
Central nervous system damage without motor involvement		8
Normal children—slow development		3
Miscellaneous		3
Total		131

TABLE II. SECONDARY DIAGNOSIS IN SAME 131 CASES

Mental retardation		39
Strabismus		30
Serious eye or visual disturbance		16
Blind	3	
Enucleated eye	1	
Hemanopsia, cortical visual disturbance, etc.	12	
Seizures		17
Speech disturbances, including so-called aphasia		16
Deafness—total		1
Multiple congenital anomalies		13
Congenital heart		5
Congenital or aplastic hips		5
Psychological problems, including autistic and so-called schizophrenia		17
Microcephaly		7
Arrested hydrocephalus		4
Total		131

TABLE III. ETIOLOGICAL FACTORS

Congenital anomalies	30
Complications of pregnancy and delivery at term	25
Prematurity per se and/or with complications	23
Postencephalitis	6
Viral infections, first trimester	5
Kernicterus	4
Twin births with complications	4
Injury—postnatal skull fracture	3
Total	100

PHYSICAL STATUS

The Iowa growth curves were used for evaluation of nutrition. Of ninety-nine children studied by this method 73+ per cent fell on or below the 3 percentile line in height, and 70+ per cent below it in weight. In twenty-two cannot readily be explained on the basis of nonuse or the failure to exercise normally. Many children with a normal bone age were much more severely handicapped so far as motor activity was concerned.

The teeth, in general, erupted in a fairly normal time and manner, even in the children having a retarded bone age. In the severely spastic, and frequently in the athetoid, the teeth were found to be ground down almost to the gums, and the oral cavity was deformed, usually with a high, narrow palate and a marked overbite. This presumably occurs from the involuntary muscle pulls.

In a sampling of blood counts, the hemoglobin was well maintained. Undoubtedly this is due to the slow weight gain which does not put particular stress on the iron storage.

The picture presented by these children, therefore, is that of small and fragile individuals. They have a poor appetite, vomit frequently; they have many respiratory infections; they perspire a great deal, making the care of the skin particularly difficult. To date, four deaths have occurred in the group. The children recall the experiments of Duraiswami[4] on chick embryos treated with insulin. He describes the chicks as delicate, fragile, and hard to rear. Studies in schizophrenia like those carried out in the Tulane department of psychiatry and neurology[5] would indicate that the major reason for the fragile constitution of these children, as well as the specific pathology such as retarded bone age, are neurogenic in origin. It is tempting to explain the physical retardation on poor appetite, difficulty in eating and swallowing, and various other handicaps, but it is in all probability central in origin and not due to these secondary factors per se. When biochemical and electrophoretic studies are carried out, probably additional constitutional variations on the same basis will be discovered.

INTELLECTUAL STATUS

That brain damage should frequently interfere with normal mental and intellectual development is axiomatic. The severity may depend on the type, extent, and localization of the lesion or lesions. Intelligence tests, modified for the handicapped, have been the subject of much research. The younger the child, the more difficult is the evaluation for the obvious reason that development early is based almost entirely on physical skills of which the brain-damaged child may be totally or partially deprived. Often the educator can better estimate the educability of the child than can the psychologist and the two disciplines should work closely together, as well as with the physician and members of other disciplines such as physiotherapy, occupational therapy, and speech therapy, in order to obtain as accurate an estimate as possible of the child's intellectual capacities. The necessity for early labeling of brain-damaged children with an I.Q. is being abandoned in favor of other methods of interpreting development and capacity. The snap diagnoses of mental deficiency by physicians are still too frequently practiced and often justifies the rage of parents of handicapped children toward professional personnel. Random examples suffice to illustrate the errors made. We have found that a child can vary in I.Q. from 70 to 105 in one year. An athetoid child rated as mentally retarded by competent psychiatric methods at the age of 18 months, at 4 years shows superior ability. In one instance, a child seen by the same group on two successive years was thought to be "obviously retarded" in the first instance and "obviously superior" in the second.

Brain-damaged children follow the same general developmental pattern that normal children do, although they may do it at a slower pace. The 2-year-old is negativistic, dependent on his mother, and individualistic in his play. The same child at 3 is more eager to enter into play with the group; he has graduated somewhat from his dependence and is far more cooperative. He may suddenly show

skills that it had been assumed were beyond his capacity. There is no justification for condemning these children during the first five years of life to the classification of mental defectives. It is better to expose them to every opportunity and stimulus possible and judge their capacities later. If this is not done, the unfavorable environment created by pessimistic attitudes may be responsible for failure to attain inherent potentialities. It should also be noted that in our experience children with pneumoencephalograms showing cortical atrophy early in life may later have normal studies and show surprisingly good development.

EMOTIONAL STATUS

It is generally accepted that human beings have a threshold to stress and strain that varies greatly, but no one is invulnerable to a breaking point. Our military personnel probably furnished the best recent examples. It is assumed, though undoubtedly erroneously so, that infants are immune to similar breaks under stress. The emotional environment that these children are subjected to are exceedingly pathological. Frequently the most tender parents forget to play with the child and talk to him, simply because the child is incapable of responding in the customary manner. In addition, the child is often hospitalized and painful procedures are carried out. Children have pneumoencephalograms, craniotomies, eye operations, feet or hip casts, and numerous other, often multiple, such experiences. It is reasonable to wonder how much this may contribute to mutism, to autism, or to the schizoid type of child one sees in the group. Less attention to I.Q.'s and more to healthy environment should be the concern of the personnel in contact with these children, especially so during the first five years of life. Their emotions certainly call for as careful nursing as their physiques.

SOCIOECONOMIC PROBLEMS

The handicapped child usually presents the parents with a financial burden which cannot be met except possibly by the very wealthy. The multiple handicap nature of the disease requires not only one consultation, but repeated visits to many doctors and other professional personnel. In addition to the pediatrician, he will usually be seen by an orthopedist, a neurologist, a neurosurgeon, or both, an ophthalmologist, frequently by an otologist and speech therapist. More than average hospitalization is required, and expensive procedures such as electroencephalograms, x-rays, and braces are usually necessary. Even care in the home is greater and often more expensive than for the normal child. Special baby sitters at a higher rate are often indicated. The mother is never as free to take the child out in the car or in the stroller as she is with the normal. Everything about him adds to the expense of care. Frequently, insurance is not available to these children. The families are in need of a realistic approach toward help in meeting the real and justifiable demands of the handicapped child. The numerous organizations, parent sponsored, that are appearing is evidence of the reality of this need.

THERAPY APPROACH

The first essential for therapy is an accurate medical diagnosis. Cerebral palsy almost more than any other

disease requires teamwork. In this team the pediatrician should be in charge of coordinating the diagnostic and therapeutic programs and interpreting the results to the parents. It is important to establish as nearly as possible the site and type of the main lesion as well as the functional by-products of this. The motor handicap is usually the most readily determined, the others are more difficult. If vision is not normal, how is it affected? Is it ocular or central or both in origin? Hearing may be present, but is the range normal? Is there present any type of aphasia? What is the child's real intellectual capacity? Is it obscured by either his physical or his emotional handicap? After the diagnostic study, it is necessary to have a realistic working estimate of the child's ability. Roughly, this can be classified as (1) possible restoration to normal, (2) limited, but self-sufficient eventually, (3) partly dependent always, (4) probably totally dependent always. This should be a working classification, and reevaluated at intervals of three, six, or twelve months, depending on the age of the patient and the progress made. After the child has been thus classified, all rehabilitation efforts should be directed toward the goal. In the young child, primarily under 3 or 3½, much of therapy must be done in a play situation. This is the great advantage of the nursery school, for in addition to being able to accomplish exercises and corrective measures, the child benefits tremendously socially from this group work. Probably, finally, socialization will be the greatest therapy the child has experienced of practical value for his future development. This is particularly true if parent education is accomplished at the same time and family adjustment can be established.

SUMMARY

From a pediatric point of view, brain-damaged children are small, fragile, and subject to frequent infections.

The intellectual capacity of these children is difficult to determine because of physical handicaps. Final judgment should be postponed and the parents encouraged and instructed in techniques to stimulate the interest of the child.

The children should be guarded against unnecessary traumatic procedures and separation from mother even more than normal children, lest such exposure superimpose on the child psychopathological damage.

The cost of adequate care of the brain-damaged child is great and frequently cannot be adequately met by private means. Diagnostic studies and therapy should be done by qualified teams and every effort should be made to rehabilitate the child and to support the parents in their attempt to do this.

The nursery school situation is a valuable therapeutic adjunct to the life of the young handicapped child.

REFERENCES

1. Thelander, H. E.: A Preschool Cerebral Palsy Program, J. Am. M. Women's A. 9: 157, 1954.
2. Thelander, H. E.: Childhood Ecology Seminar, J. PEDIAT. 44: 304, 1954.
3. Thelander, H. E.: Childhood Ecology. Factors Influencing Maturation, California Med. 81: 314, 1954.
4. Duraiswami, P. K.: Insulin Induced Skeletal Abnormalities in Developing Chickens, Brit. M. J. 2: 384, 1950.
5. Studies in Schizophrenia, Tulane Department of Psychiatry and Neurology, Harvard University Press.

Pediatric Management of Brain-Damaged Children

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A previous study showed a high percentage of children with brain damage to be small, fragile, prone to infection and difficult to rear. [4] In this presentation will be discussed more fully measures taken to help these children and their parents to make that rearing a little less difficult. It should be axiomatic that brain-damaged children, regardless of the severity of mental retardation, are entitled to accepted practices of pediatric supervision and to immunization.

PEDIATRIC MANAGEMENT OF THE MULTIPLE HANDICAP PROBLEM

The original analysis of our children revealed that they averaged 2.3 major diagnoses per child. The most common ones were visual defects, seizures and mental retardation. Deafness was not frequent, but abnormalities in hearing or sound perception occur often enough to alert us to the need for careful study by the best modern techniques in order to interpret hearing defects correctly. Closely allied to hearing are speech problems which may be due to partial deafness or to disturbed perception. Muteness or aphasia-like manifestations are relatively common.

The pediatrician, therefore, needs help from other specialists, both medical and ancillary. Practically every child should have an eye examination, even if there is no obvious evidence of abnormality. Very often hearing tests are indicated, and these should always be done if there is disturbance in speech or in interpreting speech and on children who apparently pay no attention to commands or to their name being called. Neurological and orthopedic examinations are often necessary or helpful. Psychological evaluation, to help determine the mental capacity and to interpret behavior, is especially important and will be discussed separately. Help from speech therapists, physical therapists and occupational therapists is usually necessary. It remains for the pediatrician to put all the findings together, to plan the course of therapy with the consultants, to interpret the findings and treatment to the parents and to assume responsibility for re-evaluation of the child and the therapy from time to time, usually every three to six months.

NUTRITION AND ROUTINE CARE

In our series over 70% of the children were on or below the three percentile line in height and weight as reported in a study made at Children's Medical Center, Boston, Mass. This stunting in development is primarily due to pathology in the central nervous system. There are, however, contributing factors to malnutrition. Many of the children have difficulty in eating because of the poor control of the muscles of mastication. Vomiting is a common symptom, and constipation is frequent. The nutrition of these children can be improved by careful attention to the diet, supplying one well balanced in nutrient and vitamins and easily masticated. Patience in feeding is essential. The attitude should be one of comfortable encouragement both in the management of solid food and in self-help, without anxiety or necessarily conformance to the usual routines. Frequent small feedings may be better tolerated than the usual three meals a day. Although iron deficiency anemia may occur, this has not been a major problem in our group, probably, because of the slow weight gain.

Dental care presents a difficult problem. The teeth are often ground down to the gums. Caries occur, and distortions due to abnormal muscle contractions pose an orthodontic dilemma. A general anesthetic with a careful survey of the dental needs and a repair of caries is often the only method that can be used.

Many physicians do not immunize these children due to fear of adverse reaction resulting in greater brain deterioration. Such fear, in general, is not justified. It is well to give injections with a degree of caution, starting with half the usual dosage and noting any reactions. If a child is subject to seizures, an increase in the anticonvulsant drug during the febrile period of a smallpox reaction is a wise precaution. Injections should not be given during periods of infection or diarrhea.

THE MANAGEMENT OF INFECTIONS

Brain-damaged children are fragile and prone to infection. Part of general therapy consists of attendance at nursery school which increases exposure. If there has been a definite source of trouble such as diseased tonsils, there has been no hesitancy on our part in performing a tonsillectomy. Recently two of the children have showed marked improvement after adeno tonsillectomy.

The prophylactic use of antibiotics was tried last year, using Bicillin long-acting tablets of 200,000 units each which were supplied by Wyeth Laboratories. Prophylaxis has had a long trial period in rheumatic fever, and it has been fairly conclusively shown that antibiotics in small daily doses reduce recurrent attacks of this disease. In rheumatic fever, however, the infecting agent, the streptococcus, is known and is sensitive to penicillin. In a general condition such as the symptoms produced by a brain lesion, no such specificity exists. On the other hand, Rantz has noted that in very young children, infections due to streptococcus and pneumonococcus do not produce the signs and symptoms that they do in older children and adults. [3] Often the only manifestations are rhinitis, fever and malaise. Furthermore, viral infections, uncomplicated by bacterial invaders, are self-limited and relatively short. We therefore felt justified in trying a prophylaxis to reduce the number and severity of infections. Eighteen of our children were given one Bicillin tablet daily during the past school year. The majority of the children in the nursery school who were on the prophylactic dose were maintained in apparent better health than the previous year. The parents were enthusiastic to the point of using the prophylaxis on normal siblings and purchasing the drug when the free supply was exhausted. Prophylaxis is not advocated as a routine measure but may serve a very real purpose over certain difficult periods.

SEIZURES

In our reported series 19% of the children were on therapy for seizures. [4] An additional number were and are prone to convulsions with a febrile illness. In infancy and early childhood slight seizures may go unrecognized by either parents or the physician. The children are often very sensitive to noises, the startle reflex is easily provoked, they may have tremors, ataxic and athetoid movements that mask central discharges of a convulsive nature. Some of these are so rhythmic, frequent and slight that they suggest flicker discharges. The children should be observed intently for any such symptoms and, if

present or suspected, should be tried on anticonvulsant drugs. Control of mild, frequent seizures may give the child more comfort and also improve his ability in acquiring new motor skills. Electro-encephalography may be of value in evaluating susceptibility to convulsions.

The common anticonvulsant drugs are tried first. There are many of these now available, most of them not unduly toxic for children. For major seizures, phenobarbital, mebaral, dilantin, mesantoin, mysoline and phenurone are used. For petit mal attacks, benzedrine or dexedrine, tridione and paradione are tried first. Any combination may be resorted to in difficult cases. Dilantin has produced toxic symptoms, usually an ataxia that may be confused with the original central nervous system disease. It should be given with some caution. Fairly frequent blood counts are indicated, especially when a drug is first used.

TRANQUILIZING DRUGS

The tranquilizing drugs, Serpasil, Chlorpromezine and Equanil, have been tried on selected cases the past year. The twenty children placed on Serpasil, which was supplied by Ciba Company, fell into three groups: (1) Children with atypical seizures not satisfactorily controlled by the usual drugs; (2) Children with autism as part of the symptom complex; and (3) Children with distractibility and hyperactivity. There were very few adverse effects from the drug administered in dosages of 0.25 mgs. three times daily. A few children had a temporary rhinitis, two became so drowsy that the therapy was discontinued and one child was nauseated by it. There was no evidence that seizures were helped by the drug. There was no improvement in muscle control or function. Five of the 20 children, all in the group of hyperactive and distractible, were more easily managed and more trainable. Chlorpromazine was more effective than Serpasil in two children. Equanil has had a quieting effect on one child and relaxed rigidity somewhat in another. It has caused more nausea to date than either of the other two drugs. This is a preliminary survey of the use of these drugs in brain damaged children and, while trying them, we also await reports from other centers.

PSYCHOLOGICAL PROBLEMS

Crippling is fraught with emotional problems for the child and the family. At the time of recognition, the parents need the main support. Interpretation of the condition should be done only after complete study and evaluation of the child and his potentials, using the skills of medical and ancillary colleagues. There are on record errors by competent physicians in which children diagnosed as mentally deficient have grown up to show not only great talent but ability to conquer the handicap satisfactorily. Two books written by patients should be familiar to physician and parents alike, namely **Born That Way** by Carlson and **My Left Foot** by Brown. [2, 1] These success stories, on the other hand, often raise the hopes of parents unduly. The physician has a great responsibility in guiding parents to accept the child's abilities realistically and helping them plan the future accordingly.

TRAINING AND EDUCATION

For the child with a severe motor handicap, just as for the child who is blind or deaf, the environment must supply the experiences that the normal child seeks and attains by himself. We are just beginning to understand the importance to normal maturation of such simple experiences in infancy as mouthing, manipulation of objects, feeling and exploring.

The aims in the nursery school are: (1) To give these children as many experiences of normal children as possible, that is, to see, touch, feel, explore, hear and smell the world around them; (2) To be with other children, see and share with them; and (3) To be accepted by adults as individuals, not as objects of pity or aversion. Motivation plays an important role in accomplishment. Acceptance minimizes self-consciousness and makes muscle control, speech and performance easier. The school gives parents an opportunity to evaluate realistically the areas in which the child can hope to perform and, therefore, discourages the needless pursuit of unattainable goals. The staff helps to interpret to parents the child's readiness for certain skills. Parents are often uninformed, especially with a first or an only child, as to the age at which normal children are ready for various skills, such as talking, feeding self and training.

Our facility is designed primarily to work with the preschool child. One function, therefore, is to prepare the children for more advanced work and to help parents in the selection of the school or training program. In larger urban communities, there is a great variety of facilities but this may not be the case in smaller cities and rural areas. One aim is to enter as many as possible of the children into the normal school programs. This is important both for the handicapped child who must live and work with normal people and for the other children who need to see that even with a handicap many normal functions are possible. Others go to special schools for the handicapped, and the most severely involved may do best in day care homes where they get some therapy and limited training. Those who are mentally retarded as well as physically handicapped may need hospitalization in state institutions.

SOCIAL SERVICE

The social worker is of great help to parents and to the staff. Planning for these children calls for knowledge of all the facilities in the community and for sensitivity to the parents' reactions to personnel, to other patients and to the environs of the institution. Education and helping parents to accept the facts gradually is important. Parent groups, or even waiting room conversations, increase the understanding of parents and help them to adjust to the problem.

SUMMARY

A review of the pediatric management of children attending our cerebral palsy clinic and nursery school is presented. The integration of many disciplines in order to diagnose correctly and plan therapy is one of the utmost importance. The pediatric management of nutrition, immunization, control of infections and control of seizures is briefly outlined. The emotional problems of the children and parents are reviewed and the educational problems posed are discussed.

BIBLIOGRAPHY

1. Brown, Christy, *My Left Foot*, London: Decker and Warburg, 1954.
2. Carlson, Earl R., *Born That Way*, New York, N. Y.: John Day Co.
3. Rantz, Lowell A., Maroney, Margaret, Di Caprio, Joseph M., "Hemolytic Streptococcal Infection in Childhood," *Pediatrics*, XII:5, November, 1953, 498-515.
4. Thelander, H. E., "Pediatric Evaluation of Brain-Damaged Children," *Journal of Pediatrics*, XLVI: 6, June, 1955, 699-703.

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Children, the Victims of Fragmentation Of Health Services

Children are important. They are the men and women of tomorrow. Their physical, mental, and emotional well-being is the responsibility of the medical and allied professions. There is a great deal of money, public (federal, state, and local) and private, (a charity fund for every day of the year) expended and yet it is practically impossible to finance adequately simple programs of child care. The delinquency rate, the neurotic and psychotic statistics, indicate that such programming is important and should be supported.

At the end of World War II the Academy of Pediatrics sponsored a nationwide study of health services for children. I participated in the study and for several years thereafter attended national committee meetings concerned with medical education leading to better child health care. For the past 10 to 15 years as Chief of Pediatrics at Children's Hospital I have tried to analyze the nearly insurmountable problems encountered in programming for children in need of special help. This communication is an attempt to record the findings of this study. Three different groups of children will be briefly reviewed: (1) The handicapped child, (2) the deprived child, and (3) the health problems of the normal child.

The Handicapped Child

Children with a chronic or longterm handicap such as central nervous system disease, blindness, deafness, cardiac anomalies, metabolic diseases, congenital defects, etc., need planning, as follows: (a) diagnosis, (b) therapy, (c) education, and (d) counseling. The planning and staffing for this type of program is not difficult nor unduly expensive, especially if the preventive aspects of such programming are considered. The financing and the execution of the program is well nigh impossible. Diagram 1 is an attempt to clarify the reasons. Money for diagnosis of some diseases can be obtained through Crippled Children Services. The C.C.S., however, has grown haphazardly and new diseases have been added without overall planning. This leads to some difficulty. The Crippled Children Services should be reviewed and the diseases coming under their jurisdiction reevaluated. This should not be too difficult a procedure and certainly calls for pediatric participation in any such reevaluation.

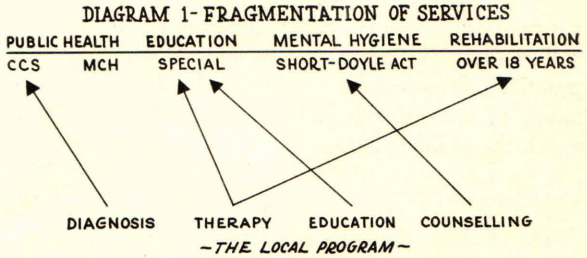
Diagnosis, however, is the easiest part of the problem. When therapy is contemplated the Crippled Children Services give very little direct assistance except for surgical procedures. The daily therapy, for instance, of the cerebral palsy child is administered only through the school system and the financing comes down through the Department of Education. Crippled Children Services can participate in therapy for the cerebral palsy child only through this system. The clinics for the cerebral palsy child are set up in the School Department. Originally these clinics were conducted by orthopedists and to a large extent they still are so conducted. If a patient has minimal motor involvement but a learning problem such as an expressive aphasia, he is not eligible for such a clinic. If he is too handicapped to be educable, he has no choice for therapy except through the school clinic. As the crippling problems became recognized earlier in the child's life, schooling was pushed back to accommodate the kindergarten children and then the nursery school children. Now that these conditions are often recognized at birth or shortly thereafter, these babies have to go to the school clinic to get any help and usually the type of help that is given in the school clinic is not either what the child or the parents need.

Legislation for day care for the severely handicapped uneducable child is also in the hands of the Department of Education. This group of children need nursing care as they are untrained, they are not educable and usually not trainable, they are subject to convulsions. Physiotherapy and occupational therapy will help them some but mostly they present a problem of day custodial care. It is "baby-sitting" and educators should spend their time at something besides baby-sitting. These day care centers should be under pediatric supervision and associated with hospitals.

The education and training of the handicapped child is also exclusively in the hands of the educators. There are state schools for cerebral palsy, the blind, and the deaf; there are local schools for children with motor handicaps; there are schools and classes for the deaf, the blind, and the retarded. The function of each of these schools is clearly defined and limited. The admission, retention, or expulsion of a pupil is under the control exclusively of the educators.

A handicapped child rarely escapes with only one handicap. There is no provision made for a cerebral

DIAGRAM 1- FRAGMENTATION OF SERVICES



CCS = CRIPPLED CHILDREN SERVICES.
MCH = MATERNAL CHILD HEALTH.

palsy child who is blind and/or deaf. The deaf school will not take him; the blind school will not; and the cerebral palsy school will not accept him. The same applies to a hyperkinetic child with or without retardation. A similar policy is practiced in the state school for these handicaps.

A handicapped child needs repeated evaluation and the parents need counseling. This calls for the services of psychologists and social workers. State money for such services comes through the Mental Hygiene Division, more particularly the Short-Doyle Act. Again, these funds are so delimited and their use so defined that it has been impossible to utilize these monies for help for either the handicapped or retarded in the program.

The rehabilitation services of the state, again a separate department, are limited primarily to adults over 18.

The private charity fund drives raise a tremendous amount of money and our people are willing to give to these funds. Again, each charity clearly defines its function. It will support only its own specific disease and only in its own prescribed manner. The result is that children in need of help either fall between the charities or their heads fall in one classification and their feet in another. The charity table is heaped with good intentions but the children and the programs aimed at helping these children get only the crumbs that fall from this table.

A possible solution to the above fragmentation at the state level would be removal of the children from all of these divisions and the creation of a special department for the handicapped child. This naturally should be in the Department of Health with a pediatrician in charge. Members of the other divisions, namely, Education, Mental Hygiene, and Rehabilitation, should work in close cooperation but be ancillary to the pediatrician in charge of such a division.

At the local level the programming and habilitation for these services should be in hospitals, again, under pediatric supervision. The clinics should be removed from schools where they do not belong and placed in hospital settings.

Those in charge of the charity funds should make a determined effort to look at the children and their needs and support these programs accordingly rather than by the sharp delineation which is now practiced. It would be timely also to determine how much of these funds are "consumed" before they reach the children.

The Deprived Child

The Public Welfare Department has a division known as Aid to Needy Children (A.N.C.). This aid, however, includes the parents, primarily the mother, of the needy child. The medical program, probably in part to economize by utilizing existing facilities but also possibly in order to please the private physician, was set up as follows: (1) Well child care was to be obtained through the existing well baby clinics; (2) hospitalization, when necessary, was delegated to the county hospital; (3) private physicians would be paid for making home calls on the sick. The result of this program is a discontinuity of care that baffles the patient and has completely discouraged the physician. The private physician can treat an A.N.C. child for diabetes but if that child gets a severe infection or goes into diabetic coma he is sent to the county hospital to be treated by a new set of doctors. Pediatricians have told tales of how after a diagnosis of acute appendicitis or pneumonia the child was sent to the county hospital only to be sent home with the statement that he was eligible for private care. These tales are innumerable.

In order to eliminate some of this fragmentation a group of pediatricians, after meeting and discussing the problem, suggested the following, namely, that well child and routine care be in existing hospital clinics, including the well baby clinic in the hospital. This same hospital should then be utilized for any illnesses of the patients registered with them. A group of staff physicians would volunteer to make home calls on patients so registered. This suggestion has been sent to the local county medical society and to the community chest.

The Normal Child

Practically all specialties in medicine see children in some capacity and yet most of the physicians in specialties other than pediatrics have had little or no training in childhood diseases or with children per se beyond that given in undergraduate medicine. There is now a tendency to have straight internships and even in many rotating internships pediatrics is reduced to one month. This lack of training in the characteristics of the young has serious implications. The toxicity of drugs is not sufficiently tested for the young with consequences such as blindness from excess oxygen, gray baby deaths from chloromycetin, etc.

The hazards of radiation have been given a little more attention but this is still largely in the "lip service" stage.

Although there is an increasing number of pediatricians in the subspecialties such as allergy, metabolic diseases, cardiology, hematology, etc., these conditions are still often handled by internists who know a great deal about the disease but very little about the child, his growth, or his ego needs.

The surgical specialties are the greatest offenders. These add to the hazards of medication and emotional trauma those of anesthesia and hypoxia to the growing brain. Frequently also, because of lack of

knowledge of the child, water intoxication occurs or excess blood is given. The pediatric surgeon has had a formidable struggle indeed to get recognition.

Since children are our greatest asset, the medical school should not end the study of growth of human beings with the completion of the course in embryology. Growth, development, maturation, aging, and ecology should be a basic course in the medical school continuous through the undergraduate years. Every specialty and subspecialty that deals even remotely with children should be required by their respective boards to have an intensive course in pediatrics before certification.

Although this communication deals with children, the same situation probably exists to a varying degree in all of medicine and the allied services. Is it not opportune that as physicians we take some active steps to remedy it?

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LETTERS *to the Editor*

The Fetus and Its Environment

The excellent work of the California Maternal Mortality Study Committee reported by Leon Parrish Fox, M.D., in the CMA of September prompted me to pull out 100 records at random from our files of children born with defects in order to review the prenatal histories. Our figures refute the oft stated belief that causes of defects are not known in most instances.

Of the 100 cases—there were 3 adopted children in which the histories were not known and there were 3 cases in which the pregnancy, delivery and family history appeared normal. This left 94 cases adequately explained. Of the total group 13 per cent had either hypoxia or trauma at birth; 16 per cent had genetic defects or a family history of birth defects. In 34 per cent there were maternal health factors adverse to normal fetal development and in 31 per cent there were unfavorable intrauterine factors. The maternal factors included infections early in pregnancy, drugs, radiation, toxemia, etc. Two striking factors appeared; 6 per cent of the mothers were outside the optimum child bearing period, over 39 or 40 years or under 16 years, and there were 5 older mothers to one younger (15 years). Fifteen per cent of the mothers had a history of psychosis before or during pregnancy, or a severe shock early in pregnancy (such as seeing one of her children hit by an automobile) or prolonged emotional stress and unhappiness (as one mother said, "I cried for nine months").

Of the intrauterine factors the most common cause was bleeding. Usually this occurred early in pregnancy but it was also associated with placenta praevia and premature separation. Other factors included multiple births, cord entanglements, RH incompatability and pre- and post-maturity. When the prenatal histories were considered instead of birth weights, it was found that the same factors were operative in both children born prematurely and at term. In other words bleeding early in pregnancy might cause defects and *premature delivery of the fetus* or it might cause defects but the fetus was carried to term. This casts a new light in the subsequent development of premature infants.

The pregnancy study in Kuhai reported by Dr. Bierman* and staff has been reviewed and it will be noted in her figures that for every prenatal death there were two surviving severely handicapped children (there were more than two if lesser degrees were included). The residium of crippled and retarded children and maternal and fetal mortality are interrelated. This points out the necessity for Obstetrics, Pediatrics, Public Health and other health services to combine their efforts to reduce the chain of events that lead to maternal and fetal mortality and morbidity.

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*Bierman, Jessie M. *et al.*, The community impact of handicaps of prenatal or natal origin, Public Health Reports, 78:10, Oct. 1963.

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CONSEQUENCES OF NEONATAL PATHOLOGY

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CURRENT interest in cerebral palsy has led several investigators to make careful studies of prenatal and birth histories of brain-damaged children. Most of these have been retrograde studies. In order to get a better perspective of what actually happens when newborn and young infants are victims of disease and injury, three different groups of babies have been investigated at Children's Hospital in San Francisco. Two of these are follow-up studies and one is retrograde, as follows: (1) Infants from the newborn nurseries who at birth or during the hospital stay had symptoms or signs that might indicate central nervous system injury or disease. (2) Newborn and young infants admitted to the pediatric or contagious disease units with disease that might produce damage to the central nervous system. (3) Children in the cerebral palsy clinic born at the hospital and whose newborn records are comparable with the above group (1).

MATERIAL

Children's Hospital is a general hospital admitting primarily women and children. In addition to the usual departments, it has a contagious disease unit. The newborn nurseries are under the direction of the department of pediatrics but there is a close liaison between obstetrics and pediatrics. The pediatric resident is in attendance at cesarean and abnormal deliveries. Joint discussions are held on resuscitation, replacement transfusions, and fatal cases. In a previous study[1] it was found that 90 per cent of deliveries were performed by physicians limiting practice to obstetrics. The other 10 per cent were done by a limited number of general practitioners well trained in obstetrics. Approximately 90 per cent were private patients. Since January, 1952, the resident on the nurseries has kept a file on newborn infants showing any abnormality while in the nursery. From this file was selected the material for Group I of this study. These infants now range in age from 6 months to 3½ years. It is obvious that observation over such a short period of time is insufficient. These patients require observation their entire life span. It is possible that late psychological manifestations or even longevity may be influenced by birth injury.

During the period from Jan. 1, 1952, to July 1, 1955, there were 6,687 babies delivered at the hospital (Table I). Of these, 6,610 were born alive, seventy-seven were stillborn, 460 were premature births. There were ninety-four neonatal deaths of which sixty-six were premature and twenty-eight were at term. There were fifty-five infants

From the Department of Pediatrics, Children's Hospital.

who had conditions or symptoms which might indicate future brain abnormality or deficit. This study is not designed to review the fatal cases, but two types of fatalities were deemed pertinent to this study. There were twelve deaths from erythroblastosis; eight of these were stillborn and four died shortly after birth before or in spite of transfusion; one of these latter was a premature and three were at term. There were seven deaths due to intracranial hemorrhage. Three of these were premature infants and four were at term.

GROUP I. NEWBORN INFANTS FROM THE OBSTETRICAL NURSERIES

Fifty-five newborns who might be candidates for central nervous system damage have had follow-up studies. eight had erythroblastosis, four of whom had exchange blood transfusions, and one of the others was ABO incompatibility, with no transfusion. The others had two, three, and four small nonexchange transfusions. One of these erythroblastotic infants is severely handicapped. There were five with other types of blood dyscrasia. Two were hemorrhagic disease of the newborn. One was a severe anemia believed from blood studies to be due to blood loss, probably from the cord or placenta before delivery. One patient had thrombocytopenic purpura. This infant developed extensive petechiae and ecchymosis shortly after delivery and had several convulsions. She was transfused and given cortisone. She recovered but has blindness and a moderate left hemiplegia. The

TABLE I. CHILDREN'S HOSPITAL STATISTICS, JAN. 1, 1952, TO JUNE 30, 1955

Babies delivered		6,687
Born alive (prematures—460)	6,610	
Stillborn (erythroblastotic—8)	77	
Neonatal deaths		94
At term (erythroblastotic—3)	28	
Premature (erythroblastotic—1)	66	
Neonates relative to present study		55

This was done by contacting the pediatrician or physician in charge at the time the infant was in the nursery. Most of the babies were under the care of staff physicians. On fifty-one of the fifty-five satisfactory follow-up was possible. Six patients were found to be handicapped and forty-seven had no sequelae referable to the newborn symptomatology that could be determined by pediatric evaluation. These babies are subdivided according to type of pathology in four groups (Tables II, III, IV, and V). In Table II are fourteen cases of blood dyscrasia. It was impossible to locate one of these. Of the thirteen remaining, mother was found also to have a thrombocytopenic purpura. One infant had hemolytic anemia; the type could not be satisfactorily determined. It may have been on the basis of infection. This child has impaired hearing and marked atonia. The latter two babies are in the cerebral palsy clinic. Dr. Ralph Wallerstein has made all the hematological studies on the above group.

Table III is an analysis of children sustaining head injury during delivery. There were twelve in this group and follow-up studies were obtainable on all except one. There were four known skull fractures demonstrated

TABLE II. NEONATES WITH BLOOD DYSCRASIA

TYPE	NUMBER	THERAPY		SEQUELAE		RECOVERY	UNKNOWN
Erythroblastosis fetalis	8	Exchange transfusion	4			7	
		Multiple transfusion	3	Retarded	1		
		No therapy	1				
Hemorrhagic disease of newborn	2	Transfused	2		0	2	
Icterus and anemia—type undetermined	1	Exchange transfusion	1	Deafness and atonia	1	0	
Thrombocytopenic purpura	1	Transfusion, cortisone	1	Blindness and hemiplegia	1	0	
Anemia—intrauterine hemorrhage cord or placenta	1	Transfusion	1		0	1	
Not followed	1		–		–	–	1
Total	14				3	10	1

TABLE III. NEONATES WITH HEAD TRAUMA

TYPE		SEQUELAE	RECOVERY	UNKNOWN
Skull fracture	4	0	4	
Depressed	1			
Comminuted and depressed	1			
Linear	2			
Cephalhematoma (no x-ray)	4	0	4	
Paresis	3	0	3	
Left internal strabismus	1			
Right arm	1			
Left facial	2			
Convulsions in conjunction with above paresis	3			
Subdural hematoma	1	–	–	1
Total	12	0	11	1

TABLE IV. NEONATES WITH RESPIRATORY DISTRESS

TYPE		THERAPY		SEQUELAE	RECOVERY	UNKNOWN
		OXYGEN	ANTIBIOTICS			
Anoxic—requiring resuscitation	4	4	2	0	4	
Atelectatic	6	6	5	0	6	
Pneumonia	3	3	3	0	3	
Pneumothorax	1	1	1	0	1	
Not followed	2	–	–	–	–	2
Total	16	14	11	0	14	2

TABLE V. NEONATAL DISEASE—MISCELLANEOUS GROUP

TYPE		SEQUELAE	RECOVERY
Convulsions—unknown etiology	2	1	1
Marasmic	2	1	1
Post-mortem delivery	1	0	1
Tremors, opisthotonus irritability	3	0	3
Survivor twin set (sibling macerated)	3	1	2
Survivor twin set—premature, enteritis	1	0	1
Diabetic mother	1	0	1
Total	13	3	10

TABLE VI. HOSPITAL ADMISSIONS—NEONATES AND INFANTS WITH DISEASE

TYPE		AGE	SEQUELAE		RECOVERY
Birth trauma (convulsions, skull fracture)	1	Newborn	Right hemiplegia	1	0
Survivor—macerated twin (convulsions)	1	Newborn	Quadriplegia, microcephaly	1	0
Erythroblastosis (transfusions)	1	Newborn		0	1
Convulsions—unknown etiology	3	Newborn		0	3
Severe generalized infections	2	Under 7 wk.	Severe retardation	1	1
Meningitis	3	Under 6 mo.	Retardation	2	1
Encephalitis	1	Under 6 mo.	Retardation	1	0
Total	12			6	6

by x-ray, two of which were depressed and one was comminuted. Four other infants had large cephalhematomas, but did not have x-rays taken of the skull. Three children had paresis, two of the face and one of the arm. Three of the children in the group had convulsions during the nursery period. None of the eleven followed had any nervous system sequelae, even though the one with comminuted fracture, now 3 years of age, has had craniotomy.

Table IV is an analysis of neonates having respiratory distress during the newborn period. There were sixteen infants in this group; on two no follow-up was obtained. Among the others there were four babies with intrauterine anoxia due in two instances to premature separation of the placenta, in one to premedication, and in one to long difficult labor. There were six cases of atelectasis, one of which had unilateral emphysema. There were three patients with pneumonia and one with pneumothorax. Two of the children were in the Airlock, one for two hours, and the other for seven hours. All of these infants had oxygen therapy, the time varying from one to seven days. Eleven of them had some form of antibiotic therapy. None of this group has any demonstrable evidence of central nervous system damage.

Table V consists of a miscellaneous but interesting group of babies. One infant was delivered by post-mortem cesarean section. The mother had streptococcic meningitis secondary to rupture of an old cerebral abscess. This child is normal. Two children were born at term, weighing three pounds 4 ounces, and 3 pounds 15 ounces, respectively. These both looked marasmic and in each case the placenta showed extensive infarcts. One of these children has slight hemiatrophy and curious small, fragile hands. Her mentality is normal. The other child at 3 years is normal though of small stature. Two children had convulsions of unknown etiology. One of these is normal, the other continued to have convulsions and was found later by pneumoencephalograms to have a brain defect, probably on the basis of congenital anomaly. Three children with hyperexcitability, tremors, and opisthotonus are normal. The child of a diabetic mother is normal. One very small premature infant, 1 pound 15 ounces, with bloody stools, is normal. There were three instances in which one of twins was macerated at birth. Two of these surviving twins are normal, but the other is a microcephalic, spastic quadriplegia. The latter child had no demonstrable evidence of injury in the nursery and like the other two was dismissed as normal. There were, therefore, three children in this miscellaneous group who were abnormal.

GROUP II. NEONATES AND INFANTS ADMITTED TO THE HOSPITAL WITH DISEASE

There were twelve newborn and young infants admitted to the pediatric or contagious disease wards with disturbances which might cause central nervous system damage. These are analyzed in Table VI. Six of these were admitted in the neonatal period, the remainder were admitted under 6 months of age. Of the neonates four were hospitalized for convulsions, one of these had skull fracture, cephalhematomas, and facial paresis. This

child is in the cerebral palsy clinic with a right hemiplegia. She has a normal intelligence. Another whose twin sibling was born macerated was admitted because of the occurrence of convulsions. He is now a microcephalic quadriplegia. The remaining two with convulsions of unknown etiology are normal except that one has a recurrent urinary tract infection. One child with neonatal erythroblastosis is normal. Two of the infants had severe generalized infection, sepsis, diarrhea, or pneumonia. One of these is severely retarded, the other is normal. There were three with meningitis, two of whom have central nervous system sequelae and one is normal. One child with encephalitis is severely retarded. Direct damage to the central nervous system by infection or intoxication has a serious effect in the young—50 per cent of these infants on our service have central nervous system sequelae.

GROUP III. CHILDREN IN THE CEREBRAL PALSY CLINIC BORN IN CHILDREN'S HOSPITAL

The final group analyzed were eighteen children attending the cerebral palsy clinic who were born at this hospital and whose early histories and physical examinations in the nursery afforded comparison with Group I. These eighteen patients are analyzed in Table VII. There were seven premature infants in the group; of these five were of pairs of twins, three were surviving twins, and two represented a set of twins. Of the other two premature infants, one had congenital heart disease. This infant while in the hospital and once after dismissal had prolonged apneic episodes, which attacks were believed, after careful study, to be secondary to the cardiac condition. The other one had a severe apneic attack in the nursery for which intracardiac Adrenalin was adminis-

TABLE VII. CHILDREN WITH CEREBRAL PALSY—STUDY OF BIRTH RECORDS FOR BRAIN-DAMAGE PREDICTABILITY

			PREDICTABLE IN NURSERY	NOT PREDICTABLE IN NURSERY
Blood dyscrasia		2	1	1
Injuries—trauma at birth		2	2	0
Prematures		7	3	4
Twin (one of set)	3			
Both twins	2			
Congenital heart	1			
Apnea—intracardiac stimulation	1			
Surviving twin at term (Sibling dead in utero)		1	0	1
Congenital deformities		4	3	1
Arrested hydrocephalus	1			
Hydrocephalus	1			
Strabismus only in nursery	1			
Congenital defect central nervous system	1			
(Congenital heart and premature, see above)				
Undiagnosed		2	–	2
Total		18	9	9

tered. One had beginning evidence of retrolental fibroplasia. The other four were dismissed in good condition. The pair of illegitimate twins appeared later with a history of gross neglect in the home, one having sustained a skull fracture and subdural hematoma. Of the two others dismissed apparently in good condition, one is a severe athetoid and the other is primarily a slow developer with atonic muscles and delayed bone age. One child survived whose twin sibling was macerated in utero. This infant is a microcephalic, spastic quadriplegia. There are two similar children in our cerebral palsy clinic.

Two children born at term represent sequelae of birth trauma. One of these, a severe athetoid, was a first child; the mother suffered vascular collapse with an unobtainable blood pressure while being given caudal anesthesia. Delivery was difficult. The infant was slow to breathe, had a weak, feeble cry, and several days elapsed before there was any sucking reflex. The second child was delivered after a long labor following a persistent posterior position. Like the other infant this baby had a weak, feeble cry and many days elapsed before the sucking reflex returned. This infant has cortical visual disturbance and delayed walking.

Five congenital anomalies occurred in the group. One of these died early. One has hydrocephalus. One has arrested hydrocephalus with mental retardation. One other is retarded and a slow developer. One with congenital heart disease has been described with the premature infants above. Three of these gave concern in the nursery. Of the other two, one was noted merely to have a peculiar shaped head (arrested hydrocephalus) and the other a persistent strabismus. Two of the children in the cerebral palsy clinic are the results of blood dyscrasia reported above in Group I.

There were two children in whom the cause of the central nervous system abnormality is still unknown; both of these were unrecognized as abnormal in the nursery. One is a severe athetoid and one is a retarded, autistic child.

In nine of the eighteen children (50 per cent) registered in our brain-damaged group there was no specific evidence in the nursery that they would have severe central nervous system symptoms later. In only two of these (the neglected twins) is there evidence that the damage was caused later. One infant was noted to have a persistent strabismus, but this was insufficient to interfere in the placement of the child for adoption. The possibility of a surviving twin being abnormal when the sibling died in utero is an observation made on our own material, and commented on elsewhere.[1]

DISCUSSION

The present study represents a search for the basic etiology in brain damage. It is obvious that retrograde studies of cerebral palsy alone will not give an accurate picture of neonates' reactions to different types of stress. Three facts are impressive in these tables. The first of these might be descriptively stated as, "Babies are designed to live." Newborn infants with a good prenatal history, born at term, are sturdy, robust individuals and can recuperate from considerable shock, trauma, infection, and other distress. None of the babies with head

injury or respiratory disease in the nursery had evidence of persistent brain damage at the time of the follow-up. As stated earlier, this does not preclude possible later consequences from perinatal pathology.

The second fact might be covered with the phrase, "Neonates have a past history." The present investigation and other studies reveal that premature and feeble infants often have a poor past history which indicates adverse genetic factors, isoimmunization, or an intrauterine environment unfavorable to normal development of the fetus. A maternal history of abortions, stillbirths or previous premature births may indicate a short or morbid life span for the infant that survives. Prematurity, multiple births with resultant deprivation of one or both infants of either oxygen or adequate nutrition, death in utero of one of twins, congenital anomalies, and isoimmunizations are frequently associated with brain defects. It is often impossible to separate pre- and postnatal conditions; the latter is usually related and consequential to the former. It is also impossible to separate completely one set of symptoms from another; these, too, are interrelated. Atelectasis, pneumonia, or emphysema may be secondary to anoxia in utero, to medication of the mother, trauma during delivery, or to external factors. Perinatal pathology is well reviewed in the symposium on anoxia in newborn infants.[2]

Finally, direct attack on the central nervous system of newborn or young infants by infection, or by bacterial or other toxins, results in injury in a large percentage of cases.

SUMMARY

Three groups of babies have been studied. (1) A follow-up study on fifty-one of fifty-five infants born in Children's Hospital has been made and resulted in forty-five found to be developing normally and six being handicapped. (2) Of twelve infants admitted to the hospital with diseases that might produce central nervous system sequelae, 50 per cent were found on follow-up to be handicapped. (3) The birth records of eighteen children in the cerebral palsy clinic, born at Children's Hospital, were studied. In nine, or 50 per cent, there was no evidence in the nursery that the infants would be handicapped. Prematurity, blood dyscrasias, and infection played the major role in handicapping the infants in this series. A full-term baby with a good prenatal history is a robust individual and tends to recuperate fully from the stress of being born, even when this is prolonged and traumatic. This series of cases is small; it is hoped more studies of this type will be done.

REFERENCES

1. Centano, Pilar A., Thelander, H. E., and Walters, Ruth: A Five Year Study of Prematurity, California Med. In press.
2. Anoxia of the Newborn Infant, Symposium, Oxford, 1953, Blackwell Scientific Publications.

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To the Editor:

Scant attention has been given in pediatric literature to the contributions an educator can make to the habilitation of the young handicapped child. At a recent orthopsychiatric meeting in San Francisco, Jane Phelps, the nursery school teacher in Children's Hospital Cerebral Palsy Program, discussed her observations. The paper was well received by physicians, psychiatrists, and educators alike, and several expressed the hope that it would be given wide distribution. The main points of the discussion are presented her under the headings: The Parents, The Children, and Diagnostic Observations.

THE PARENTS

"The parents require the most careful preparation for the beginning days of nursery school. The profundity of the parents' despair cannot be overestimated, for with most of our parents the diagnosis of a handicap is new. The nursery school is usually the parents' first experience in seeing a group of handicapped children together. Some children will be more handicapped than their own, some less so. The constant support the parents need in having accurate medical information repeated over and over cannot be overemphasized. The parents of a mildly spastic child may erroneously anticipate that their child will grow up to be like a severely athetoid child. Experience has taught us that we must be alert to the unspoken questions and misconceptions that are typical of parents new to the program. Here we are working with parents who are making a variety of attempts to help the child on the one hand and, in many instances, to deny the problem to themselves on the other. They may be waiting for the child 'to grow out' of it, or their expectations may be unrealistic and they may be angry and hostile or depressed and apathetic.

"We have also learned that parents may see attendance in a school situation as implying educability or trainability and such is not always the case. We therefore indicate that the first 6 weeks or so are an evaluation period in which we determine whether or not this nursery school is suitable for this child. It is basic to our discussion of what a preschool program can do for the brain-injured child to stress the fact that in this most critical preschool period one accomplishes with the child only what one is able to accomplish with the parents: helping them to accept the diagnosis and to develop an accurate picture of their child. The preschool period often marks the start of a process in which parents are just beginning to perceive the implications of the diagnosis for their child and the family unit. The birth of a handicapped child into a family is much like dropping a pebble in a pond: the effects are wide and far-reaching and affect the siblings, the grandparents, relatives, and friends of the family.

"From our experience we have learned the need for developing a flexible kind of parent education program. This may include films, speakers at monthly evening meetings, and discussion groups, but it is the direct kind of parent participation in the nursery school that enables the staff to be alert to the distortions and unasked questions that parents reveal in our day-to-day contact with them. It is here that we begin to understand the variety of attempts the parents make to 'negate' the evidence. There is tremendous advantage in being located in a medical setting where both parents and staff have easy access to the pediatrician, the psychiatrists, and the psychologists."

THE CHILDREN

"We have evolved a definite philosophy in relation to these children. We attempt to

provide simple social play directed toward emotional satisfaction in order to release whatever potential the child may have for later learning experiences. Some of the more basic satisfactions may have been restricted at home. Thumb sucking, chewing toys, feeling or smelling toys, and seemingly random, purposeless activity is not restricted unless harmful to the child or to others. It is obvious that these handicapped children are both immature and inexperienced to a greater degree than the nonhandicapped child of the same age. We share a deep concern with other disciplines as to the factors which contribute to the formation of the child's self image. It is our feeling that these children need tangible physical evidence of affection and acceptance. They need to be picked up and comforted, and picked up and played with, with expressions of acceptance verbalized when appropriate and when spontaneous. It is all too common an experience that these young children have already been exposed to reactions of revulsion, disgust, and curiosity. In the preschool years, quite apart from whatever else we can offer, if we can help them form an image of themselves as being loved, wanted, and worthy of respect, we will have contributed something basic to their development. If the basic experience here is good, we will have given them the strength to meet the stress of future contacts which may be less good.

"In this area we are reminded of various comments mothers have made to us: 'If he just didn't embarrass me by smelling everything, I could stand it better.' 'If only he didn't drool so much, I wouldn't mind taking him out.' Or, 'When my other children were 4 years old and went through a silly stage, it wasn't so bad. But when Billy does it, I'm afraid people will think he's not right in the head.'

"Because parents are already so puzzled by the odd extremes of behavior they have struggled with, they may have handled it by pressuring the child 'to stop it.' If the child's behavior is characteristically 'spotty' and swings from behavior quite appropriate to his age to behavior that seems a reversion to earlier infantile patterns, he may have had his personal dignity assaulted by 'don'ts' far more than a normal child might have to bear.

"We remember that these children may have been under medical, surgical, or psychological care for long periods of time, and may not have had much time to be what they are—young children. Whatever their chronological age, they must not be pushed into patterns of conformity beyond their maturational skills. In nursery school they are not forced to sit during snack period for longer than they are able, or forced to sit and listen to music when they should be playing.

"During the morning session the activities are much like those one would see in any nursery school although it is quieter, since many of the children do not talk; there is less moving around, since many of the children are not ambulatory, and the pace is quieter and somewhat slower. It is important that a room be available where the more typically brain-damaged child may go in order to recover his equilibrium. Such children are often conspicuous in a group, and, when pressures (which the adult cannot always see) have become intolerable, the child can be observed standing in the middle of the room with his hands over his ears, or seeking the haven of the adult's lap, or, more typically, bursting into tears. These children need a program that is individually designed for them; it will vary from day to day."

DIAGNOSTIC OBSERVATION

"During the period of unrestricted play and those periods in which some structuring is necessary, the teacher can be alert to observing the behavior, verbal responses, and problem-solving methods that offer clues as to the nature of the child's more subtle disabilities. Some of these children react to stressful situations with an increase in drooling, with unusual motor behavior such as toe walking, and an increase in awkwardness in fine finger coordination.

"One of the essential contributions a program such as ours can offer is to accumulate observed data that tends either to confirm the diagnosis or offer new insight as to the real nature of the child's problems.

"The kind of observation we can make in the playroom is a long-term process and great care should be taken to avoid placing

a label on the child that long-term observation does not confirm. We can observe the child struggling because of his inability to organize stimuli into a meaningful whole, and we can observe that he seems to interpret stimuli differently than other children. Over a long period of time we can also watch the development of avoidance techniques which these children utilize to cover up their deficiencies from the simple and direct 'I don't want to do that puzzle' to the more sophisticated 'I'm not feeling well right now.' One can watch these children struggle to remove pieces from a puzzle, expending energy in trying to remove a part that is merely a design painted on the border. The child can be observed trying to work a puzzle, but able to guide the pieces in only by placing his finger in such a way that he feels where the indentations are. We have watched such children during informal story periods, where large, colored pictures are used, and they respond in such a way that we know they are aware only of a small fragment of the total picture, or are concerned with a minor flaw in the paper which is not readily seen by the adult. Some of the children can identify the parts of the body on themselves or in a book but cannot do it on the doll. There is about these children a specific quality that suggests *interference* rather than *lack*. One must try to separate to what degree the child's anxieties and feelings of inadequacy and helplessness contribute to the formation of this symptom complex. The teacher must try to be alert to the child's changing performance to know whether his failures at present are because he has not yet developed skill in that area but is capable of so doing, or whether it may be an area which is not capable of being developed.

"Therefore, it can be said that the basic contribution a preschool program can make to the brain-injured child is to provide him with as much support and emotional security as we can to help him go on to the next learning situation. The potential of these children is often high, but, without real understanding of their basic pathology, they can be mismanaged. Then, there is superimposed a psychological problem which may make future evaluations almost impossible.

"If the first 5 to 7 years are important for the normal child, they are infinitely more important for the brain-damaged one."

JANE K. PHELPS, B.A.
H. E. THELANDER, M.D.
San Francisco, Calif.

Correspondence

To the Editor:

As a regular reader of the JOURNAL, a particular advertisement has been of some concern to me. It appears on page 79 of the June, 1959, issue. An asthenic, taut little boy is shown next to a picture of his report card marked by a teacher, indicating her "diagnosis" of anxiety. A tranquilizer is suggested for this problem. The advertisement says, in effect, to dampen all of this boy's responses to difficult problems so that he will be like other children.

Is this what we really want to achieve? The very foundation of personality development is the grappling with and the finding of a solution to these anxiety-producing situations. By use of such ataractics, are we not merely postponing, or worse, replacing this necessary agony of growing up?

The tranquilizers have been of inestimable value in medicine, and specifically in pediatrics in cases of a severe nature, but they are being used all too often to "cool off" a child in a temporary struggle with his problems. Let us not add the adolescent character disorder to our growing list of iatrogenic ailments.

EDWIN A. SUMPTER, M.D.
Nashville, Tennessee

Abnormally small head size and intellect in children

Measurements of 212 microcephalic children show them to be consistently retarded in height when compared by age and sex with 12,000 healthy children in the public schools. Abnormally small head sizes in microcephalic children have a significant correlation with intelligence quotients when degrees of microcephaly are plotted against degrees of mental retardation as measured in intelligence quotients.

Helen B. Pryor, M.D.,* Hulda Thelander, M.D.

PALO ALTO AND SAN FRANCISCO, CALIF.

STUDIES of normal head circumference for North American infants have been published by Silver and Deamer[1]; Nellhaus[2] reviewed the world literature since 1948 for Negro, Japanese, Alaskan, Eskimo, and Russian children; and Pryor[3] reported the findings in 12,000 American-born Caucasian children. Many other studies of normal children have been reported.

In his books on the dolphin, Lilly[4, 5] discusses the relationship between brain size and intellect. He used acquisition of speech as a measure of intellect and cited McCarthy's[6] study to show that a positive relationship exists between age, brain weight, and speech.

The size of the human head is determined by the growth of the brain except in cases of hydrocephalus, craniosynostosis, or other cranial pathology. Failure of the head to grow has been linked with mental retardation by a number of authors including O'Connell and associates,[7] and Book and associates.[8] Severe brain infections such as encephalitis, meningitis, or toxoplasmosis early in life may prevent further growth of the head, as reported by Thelander and Goebel.[9]

Anthropometric measurements of 181 children with Down's syndrome, reported by us,[10] revealed consistently small head circumferences at every age when compared with normal children. Their heads ceased to grow after the age of approximately 3 years, and they were short in stature.

Bailit and Whelan[11] reported an anthropometric study of body size correlated with intelligence quotients in retarded children. Compared to normal controls, the retarded children were consistently smaller.

Mosier and associates[12] studied 2,027 mentally defective patients in an institution and concluded that, for 4 main classifications, linear growth was stunted to a degree

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Table I. Age distribution to nearest birthday and ranges of head circumferences and I. Q.'s.

Age (years)	*No. cases*	*Head circumferences in standard deviations below mean for age and sex*	*No. cases*	*I. Q.*	*No. cases*
1	14	Minus 2.0- 2.9	113	100-102	4
2	34	Minus 3.0- 3.9	51	90- 99	5
3	22	Minus 4.0- 4.9	20	80- 89	14
4	26	Minus 5.0- 5.9	9	70- 79	24
5	26	Minus 6.0- 6.9	9	60- 69	22
6	24	Minus 7.0- 7.9	3	50- 59	40
7	11	Minus 8.0- 8.9	5	40- 49	32
8	10	Minus 9.0-12.1	2	30- 39	42
9	9			20- 29	29
10	7				
11	7				
12	7				
13	4				
14	2				
15-17	9				
Total	212				

Table II. Summary of clinical diagnoses

Clinical diagnoses	*No. cases*
Chromosomal aberrations	7
Inborn errors of metabolism	8
Cerebral palsy, all forms	45
Multiple congenital abnomalies, etiology unknown*	118
Postrubella, first trimester	27
Acute cerebral infections	7
Total	212

*In this group, 8 infants with syndactyly had histories of the same anomaly in other members of the families. Many others had a variety of anomalies in the families.

corresponding to the severity of their defects. We have published similar findings.[13]

Pozsonyi and Lobb[14] reported on the relationship between mental retardation and stunting of linear and skeletal growth in 958 children. Kugel and Mohr,[15] Knobloch and Pasamanick,[16] and Casalaina and associates[17] have all published data on the association of small head size and growth failure in mentally retarded children.

We report here the results of a study of the relationship of the degree of mental retardation, as measured by I. Q., to one parameter of growth, head circumference, in children with microcephaly.

MATERIALS AND METHODS

We reviewed the records of 212 children with microcephaly who came to the Birth Defects Clinic at Stanford University Medical Center or to the Child Development Center at Children's Hospital, San Francisco, during the last 3 years. In the adult, microcephaly is defined as a condition in which the skull has a capacity below 1,350 c.c. For children we use the definition that a head with a circumference 2 or more standard deviations below the mean for age and sex is microcephalic. No children with Down's syndrome are included in the present study.

In Table I are shown the age distribution, the range of cephalic circumferences, and the range of intelligence quotients for our patients.

Ages are recorded to the nearest birthday. Only 36 were 10 years of age or older. All but 2 of the 28 cases with head circumferences of more than 5 standard deviations below the mean were found in older institutionalized children sent to Sonoma State

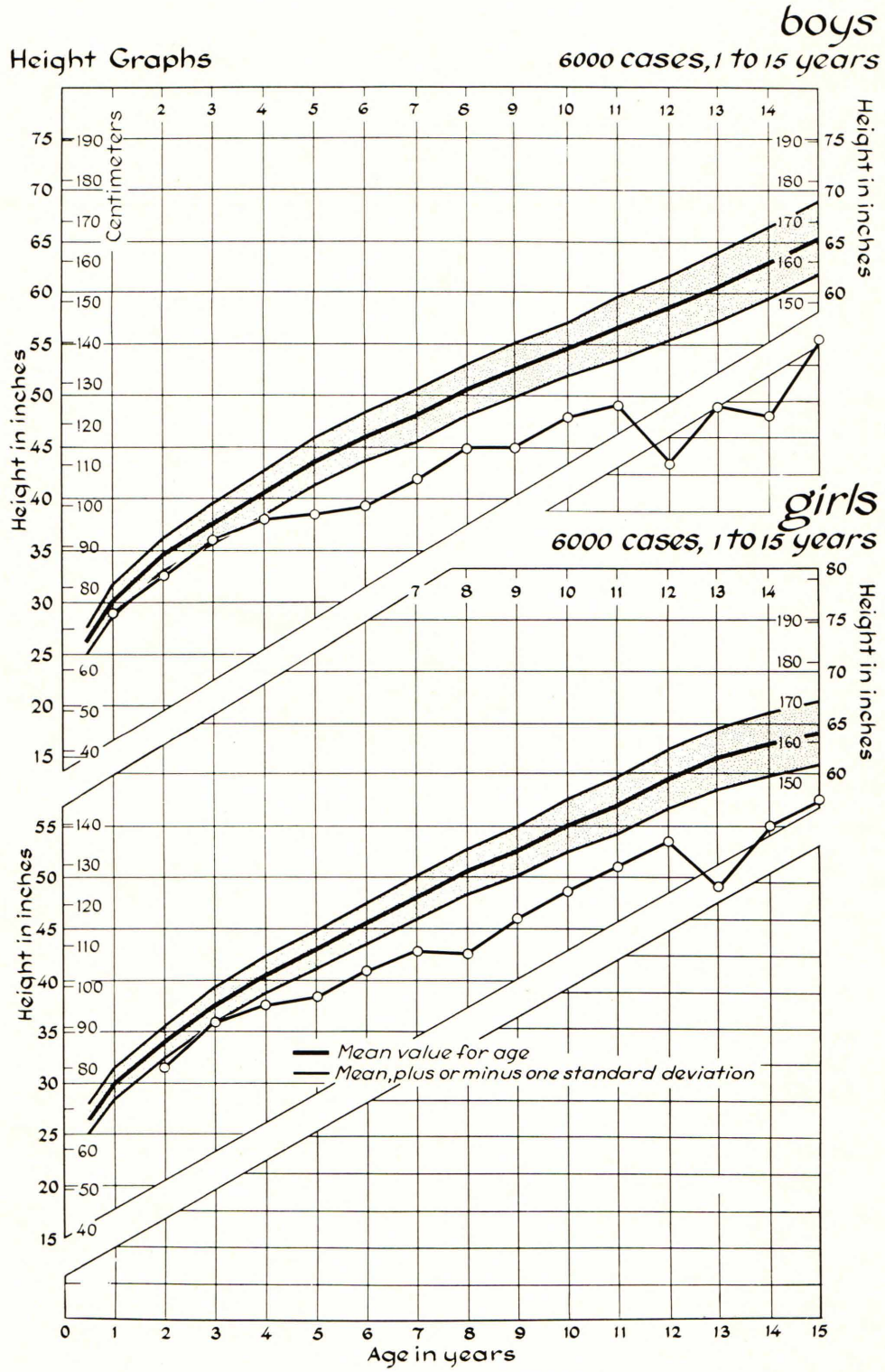

Fig. 1. Standing heights of microcephalic patients compared with normal children. Shaded areas measure plus to minus one standard deviation from the means for age and sex for normal boys and girls; O—O—O represents mean heights of 111 boys and 101 girls who were microcephalic.

Hospital, Calif. All intelligence quotients were measured by standardized, individual tests done at the two hospitals involved in the study.

Nine children, from 1 to 11 years of age, attained intelligence ratings between 90 and 102. Eight of the children in this range had cephalic circumferences from minus 2.0 to minus 2.6 standard deviations (S. D.) below the mean. The exception was a two-year-old girl with an I. Q. of 90 who had a head circumference minus 3.4 S. D. be-

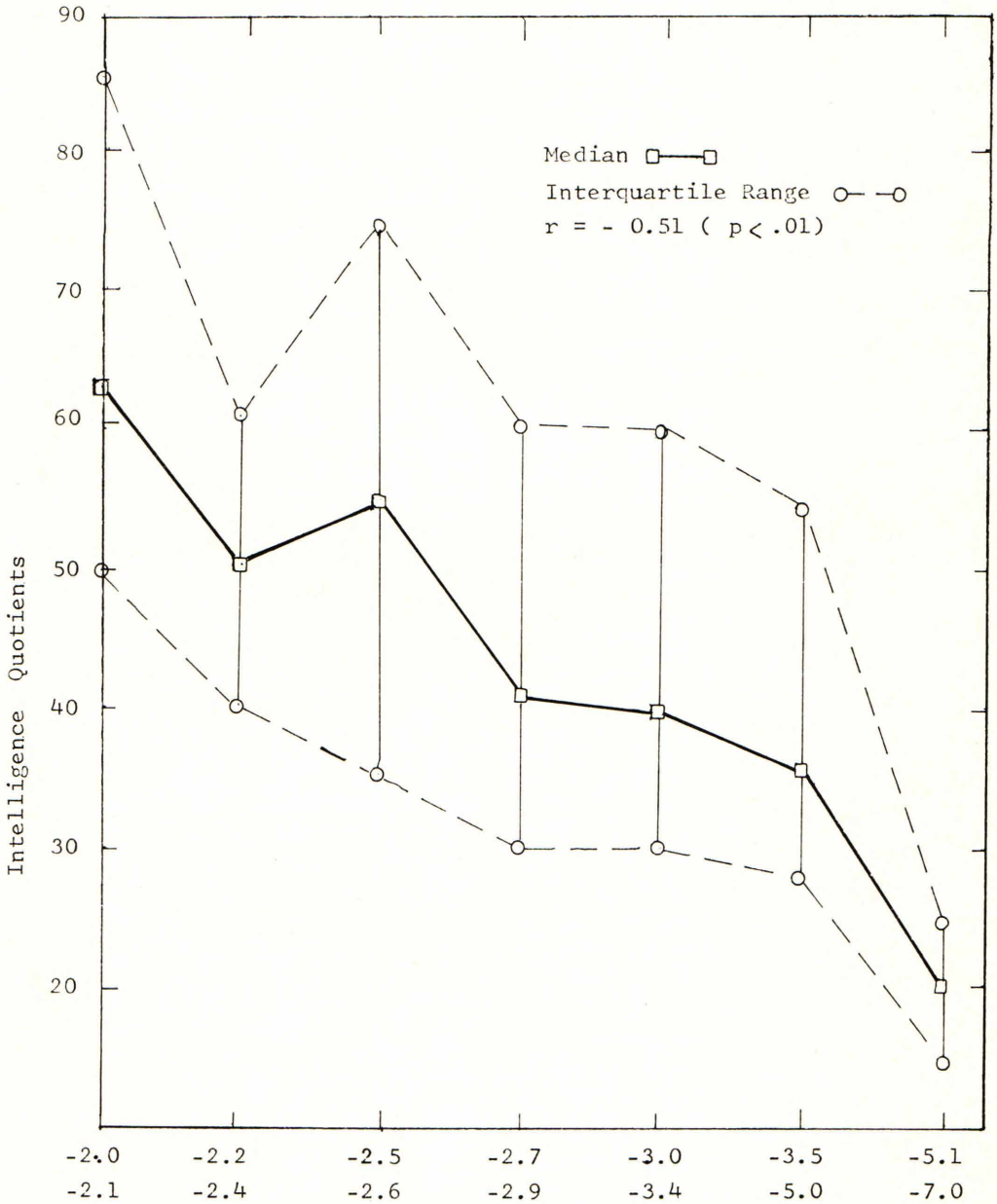

Fig. 2. Head size expressed as standard deviations below the normal means for age and sex plotted against intelligence quotients. Intervals of head circumference were chosen to include roughly the same number of observations in each group. Consequently, the intervals must be lengthened as they move away from the mean.

low the mean, and she was a congenital amputee. At the other end of the scale, all 10 microcephalic children with measurements more than minus 7.0 standard deviations had I. Q.'s of 20 or less. The 38 children with head sizes ranging from minus 4.0 to minus 7.0 S. D. had an average I. Q. of 35.6.

In the group with the top I. Q.'s, 2 children were congenital amputees, 2 had cleft palates, 2 had congenital heart defects, and 3 had cerebral palsy (hemiplegia). The latter 3 had histories of birth injuries.

Twenty-nine children had I. Q.'s below 30, and 18 of these were classified as having "primary microcephaly." Among the remainder, 2 had inborn errors of metabolism, 5 had multiple congenital anomalies, and 4 had cerebral palsy. Two of the latter had spastic quadriplegia with multiple congenital anomalies, one had ataxic paraplegia, and one had spastic diplegia with seizures. All of the group with the lowest I. Q. had more than one congenital defect. Cerebral palsy occurred among children with all grades of intelligence and all kinds of anomalies. The 3 children in the top I. Q. group, who had cerebral palsy, had no evidence of congenital anomalies or embryonic injury. Twenty-seven had histories of rubella infection before the third month of gestation. The diagnoses for the whole series are summarized in Table II. The possibility of ethnic variations was considered, but we do not have large enough numbers in the subgroups to be representative. There were 110 Caucasians, 48 American Indians, pure or mixed, 39 Negroes, 12 Orientals, and 3 of unknown parentage.

Definite stunting of growth in height is seen in our group of microcephalic children, becoming more marked as they grow older; this follows the pattern of Down's syndrome. In both groups, deficits in head size are greater than deficits in height.

In Fig. 1 the mean values for standing height of 111 microcephalic boys and 101 microcephalic girls are plotted and compared to 12,000 healthy boys and girls of the same ages in the public schools.

In Fig. 2, head size is plotted against I. Q. In the graph, medians rather than means are used for I. Q., since the data are skewed. The interquartile range is indicated by the broken lines. Since I. Q.'s below 20 could not be measured definitively, the graph ends at that point which corresponds to a head size of minus 5 standard deviations. Below this point all of our cases are institutionalized children classified as primary microcephalics. The data reveal a significant correlation coefficient of 0.51 with $p < 0.01$.

SUMMARY

Abnormally small head size has long been associated with mental deficiency. The present study relates the degree of mental retardation to one parameter of growth, cephalic circumference, in 212 cases of microcephaly. The greatest deviations from normal were found in older institutionalized children.

Intelligence quotients have a direct relationship to head size in our microcephalic patients. Some of the higher I. Q.'s were found in children whose defects could be classified as probably caused by conditions arising in pregnancy after the first trimester. The lowest I. Q.'s were found in children with chromosomal aberrations, inborn errors of metabolism, and multiple congenital anomalies. Rubella accounted for 27 cases of embryonic injury that could be ascribed to prenatal environmental insults.

REFERENCES

1. Silver, H. K., and Deamer, W. C.: Graphs of the head circumference of the normal infant, J. PEDIAT. **33:** 167, 1948.
2. Nellhaus, G.: Head circumference from birth to eighteen years, Pediatrics **41:** 106, 1968.
3. Pryor, H. B.: Charts of normal body measurements in graphic form, J. PEDIAT. 68: 615, 1966.
4. Lilly, J. C.: Man and dolphin, New York, 1962, Pyramid Publications.
5. Lilly, J. C.: Mind of the dolphin, New York, 1967, Doubleday & Company, Inc.
6. McCarthy, D.: Language development in children, *in* Manual of child psychology, New York, 1946, John Wiley & Sons, Inc.
7. O'Connell, E. J., Feldt, R. H., and Stickler, G. B.: Head circumference, mental retardation, and growth failure, Pediatrics **36:** 62, 1965.

8. Book, J. A., Schut, J. W., and Reed, S. C.: A clinical and genetic study of microcephaly, Am. J. Ment. Deficiency **57:** 642, 1952.
9. Thelander, H. E., and Goebel, G.: Microcephalia as a complication of pneumococcic meningitis, Am. J. Dis. Child. **77:** 642, 1949.
10. Thelander, H. E., and Pryor, H. B.: Abnormal patterns of growth and development in mongolism, Clin. Pediat. **5:** 493, 1966.
11. Bailit, H. L., and Whelan, M. A.: Some factors related to size and intelligence in an institutionalized mentally retarded population, J. PEDIAT. **71:** 897, 1967.
12. Mosier, H. D., Grossman, H. J., and Dingman, H. F.: Physical growth in mental defectives, Pediatrics suppl. **36:** 465, 1965.
13. Pryor, H. B., and Thelander, H. E.: Growth deviations in handicapped children, Clin. Pediat. **6:** 501, 1967.
14. Pozsonyi, J., and Lobb, H.: Growth in mentally retarded children, J. PEDIAT. **71:** 865, 1967.
15. Kugel, R. B., and Mohr, J.: Mental retardation and physical growth. Am. J. Ment. Deficiency **68:** 41, 1963.
16. Knobloch, H., and Pasamanick, B.: Mental subnormality, New England J. Med. **266:** 1045, 1962.
17. Casalaina, V., Dingman, H. F., Mosier, H. D., and Grossman, J.: The birthweight of abnormally small mental defectives, Human Biol. **36:** 229, 1964.

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Recreation as Therapy for the Retarded and Handicapped

Utilizing Recreation Productively

IN our busy and tense society, recreation usually is considered a frill. This concept may change for all of us. The Spring issue of Horizons devotes considerable space to our *new problem,* leisure. It is particularly appropriate, therefore, to recommend that we utilize recreation creatively to help those now doomed to perpetual leisure as are the severely retarded and handicapped.

The past ten to 15 years have seen a marked change in the management of the handicapped and particularly the mentally retarded. The trend now is away from institutionalization and a return to community placement. Unfortunately little has been done at the local level to meet the needs of these individuals. When interviewed, some express fear of losing companions and friends that they have enjoyed in the institution.

Doctors, largely, have been concerned with the medical aspects of diagnoses and treatment of the handicapped. Educators have tried, often in vain, to impart to the retarded the 3 R's.

Until such a time as we are more successful than we have been to date in reducing the number of defectives, we must assume responsibility for their humane care. They are human beings with the same needs as the rest of us for a home, love, friendship, fun and compassion. Behind the handicap is a child, a human being.

For the past 15 years there has been a happy working relationship between the Child Development Center and the Recreation Center for the Handicapped in San Francisco. The Recreation Center was started in 1952 by Janet Pomeroy. Janet, while attending college and working toward her degree in Recreation, volunteered to chauffeur handicapped children in the city. The combined training and experience resulted in a request to the city to use a vacant building on the beach next to the Fleishhacker pool for recreation for handicapped individuals. Six children constituted the original group; there are now approximately 500 enrolled. The ages vary from infants barely over one year old to senior citizens of undisclosed vintage.

The center is open six days a week from 9 a.m. to 9 p.m. It has a paid staff of about 60 people and more than 200 volunteers. The activities are designed to meet the needs of the different ages and also the limitations imposed by various handicaps. The young children are divided into two groups. The hyperactive retarded or disturbed children have activities which include running on the beach, walking through the woods, climbing trees, swimming, and for more quiet periods somewhat structured games. The nonambulatory youngsters play quiet games, participate in singing and mu-

sic, and go for "strolls" in wheelchairs. There are programs designed for older children, teenagers and adults. In addition to the local activities at the Center, participants are taken on sight-seeing trips around the city. Provision is made for camping out both in the city and at various resorts in the state. They even skate and ski.

The object of this report is not primarily to describe the center but to give the readers a picture of the therapeutic effect of a recreation program on the participants. The philosophy of the staff is that although a child is born less than perfect he is not therefore "sick." Behind the most grotesque deformity is a child who needs to live as nearly normally as possible. This attitude has a beneficial effect on the child. Parents are constantly amazed at the change that occurs. His appetite improves, he sleeps better, he is more self-sufficient. He begins to talk, to want to dress himself. This change is reflected in his posture, his initiative and his comfort with people.

Although the Recreation Center is not a "school," the children learn. Pediatricians know that young children learn best through play. It is only after the age of five or six years that he is ready to master the symbols of more formal learning. A retarded child who at a chronologic age of ten or 12 has a mental age of four or five still learns best through play. One object of the Center is to help the child overcome his major problems and hopefully make him acceptable in one of the many schools or classes in the city designed for children with various defects.

The older children and adults work at various crafts. Some have become sufficiently proficient to be given responsibilities around the center for which work they are paid.

The greatest benefit probably is the emotional and social adjustments that occur. A great many have been bored doing nothing at home; they have been hurt and frustrated by unfair competition with siblings and neighborhood children. The center offers them a chance to compete with peers of their own kind. They may hover around the edges at first but with music, singing and dancing they are soon enticed into the group. The transformation carries over to the home where they establish a better relationship with parents and siblings.

Not the least advantage of the Center is the partial relief it gives the mother. There is probably no situation, not even death, that is as traumatic as the birth of a child who is grossly defective. The emotional turmoil it arouses at the time of birth is followed by years of confinement and sorrow and then concern over what may happen to him should he outlive his parents.

This is a plea to physicians and other professionals in the health field to utilize personnel trained in recreation as a part of the long-term therapy for physically and mentally retarded individuals.

It is not difficult to start a recreation program. A small staff augmented by volunteers quickly become devoted workers. As an incentive to starting, I suggest the film recently released by the Recreation Center. It is called "Cast No Shadow" and portrays accurately the many activities for these children. Janet Pomeroy has authored a book called "Recreation for the Physically Handicapped," Macmillan and Company. Start!—H. E. THELANDER, M.D., *Director Emeritus, Child Development Center, Children's Hospital, San Francisco. Consultant to the Recreation Center for the Handicapped, Inc., Great Highway, Sloat Boulevard, San Francisco, California. Resident address: 40 Seafirth Place, Tiburon, California 94920.*

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Letters to the Editor

Dimensions in Cerebral Palsy Often Overlooked

SIR—Although great strides have been made in understanding the pathology of cerebral palsy there is still often an aura about the affliction that deters comprehensive interpretation of the patient.

Injury to the rapidly growing brain of the embryo, fetus, newborn and young child stunts growth and changes the subsequent pattern of development to a varying degree. Some of the setback is later compensated for but not all of it in all cases. There are several reasons for failure of the organ to recuperate.

(1) The severity of the injury. This may vary from a fatal insult to a minor one that goes unrecognized until later in childhood. Trauma during delivery may cause skull fracture, subdural hematoma, gross intracerebral bleeding, petechiae or thrombi in portions of the brain and spinal cord, or it may produce alterations in the chemistry of the child that may be as detrimental as the trauma.

(2) The design and stamina of the infant exposed to injury. Unfortunately it is often assumed that the severity of the infant's condition correlates only with the severity of the injury. The reports of psychometric evaluations and interpretation of these by the physician usually appear not to consider the type of child a given infant would have been if he had not been injured. A child with congenital malformations or hereditary disease is exposed to the same risk during delivery as the normal child. The defective child, however, is more fragile; he usually has a lower birth weight and is more vulnerable to insult than a well-designed robust infant, who also has greater recuperative powers.

There are ways of determining with some degree of accuracy the original developmental design of the child. As an example, one of the children in our unit with severe cerebral palsy is a mongol. Another child has a hereditary hemiplegia with severe mental retardation, both the mother and the grandmother were born with a hemiplegia. The assessment of a child with cerebral palsy therefore should include not only a careful family history for evidence of hereditary disease but also an equally careful history of the pregnancy and the possible insults sustained in the embryonic and fetal period. A careful examination of the newborn infant for stigmata is important. Children exposed to an adverse fetal environment may show some evidence of faulty development, even if this is not as obvious as in the babies exposed to thalidomide during the embryonic period. The stigmata that one looks for in abnormal children are such things as small deformed ears, high narrow palate, curious hair formation, simean lines, crooked fingers, overlapping toes and similar defects. These may be as important to note when doing the newborn examination as are neurological abnormalities. If, for instance, a given injury lowers the IQ by 10 points the child designed to have an IQ of 120 or 130 may compensate rather quickly from the setback and be considered normal, but the insult to a child with a potential IQ of 80 to 90 will lower his learning ability to a level where permanent dependency is the outcome.

(3) The immediate and subsequent care of the injured newborn. After a traumatic delivery some infants are in rather severe shock; others have erythroblastosis; still others suffer from loss of blood into the maternal circulation, severe hypoxia from cord strangulation or placental separation. The medical management of these emergencies is of extreme importance, though their optimum management is still uncertain. Continued observations and experimentations are indicated.

The family's emotional response to the injured child is also important. The birth of an imperfect child is a source of sorrow. If the infant has also to unfold his personality under a cloud of tears, his recuperation will be seriously hampered.

The spectrum of cerebral palsy has been extended to add children who are neurologically handicapped or have minimal cerebral dysfunction or minimal brain damage to those with severe motor disorder. The time is opportune therefore to study children with developmental abnormalities, physical and mental, both longitudinally and in profile. Individual plans for each child should be made. The present awkward method of trying to fit a handicapped child into a clinical category or a syndrome with emphasis on disease entities should be abandoned. It may be opportune to relegate the name cerebral palsy to history, along side of Little's disease. Too frequently one now finds that a neurologist takes care of epilepsy, an orthopedist manages the limb disorders of the cerebral palsy child, and special clinics are conducted for the mentally retarded. Although surgery and medication have some usefulness in these diseases these therapies are often of limited value. The far greater problem is that of rearing a handicapped child. The family and the child need the benefit of the other disciplines—psychology, education, social service and counseling. A Child Development Center, in which all these services are available to a physician and his patients, is as important a part of a Pediatric Department as the clinical laboratory. It certainly is just as important to evaluate a child's total functioning as to know his amino-acid spectrum or his chemistries.

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The Development of Human Skeletal Muscle

SIR—Dr. Fenichel begins his article in your February issue (*D.M. & C.N.* 1965, **7**, 69) with 'The mechanisms of skeletal muscle embryogenesis have been the subject of considerable controversy', but he then tells us nothing about the controversy. It is the controversial issues that are often the most important, or at least the most interesting, in any area of knowledge.

His section on histogenesis would have been much clearer with a diagram of the various structures. Regarding cytogenesis, I do not see anything particularly controversial here—even the question of whether the muscle-cell is of unicellular or multicellular origin seems to have been settled. In *The Scientific American*, Konigsberg[1] summarizes is work on cloning of embryonic muscle cells which shows conclusively that a muscle fibre can arise from a single embryological precursor cell.

On p. 71 Dr. Fenichel writes: 'A peculiarity of fetal muscle is the appearance of B fibres which are considerably larger than the others.' Although A fibres are not mentioned in the test I assume that they are the 'others'; however, one could assume the presence of many kinds of fibres. I would also assume that the B fibres are not present in adult muscle.

ANGIOGRAPHY IN A CASE OF HYDRANENCEPHALY

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ANGIOGRAPHY IN A CASE OF HYDRANENCEPHALY

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AN OPPORTUNITY for careful pre- and post-mortem studies of a case of hydranencephaly presented itself in 1951.

V. S., a female infant, was born on March 2, 1951, weighing 9 pounds 1 ounce. She was the third child of 21-year-old parents. The siblings were 3 years and 21 months. The pregnancy had been uneventful except for a feeling of fatigue attributed to repeated pregnancies and home duties. There was no history elicited of illness, bleeding, or other abnormalities in the early or later pregnancy. The baby was believed to be three weeks overdue and was delivered after a twenty-four hour labor. There was some difficulty in establishing respiration but after fifteen minutes breathing was essentially normal. The child did not take nourishment well and was tube fed for the first four days after which intake of food was satisfactory. She did not cry spontaneously. It was noted at birth that the head was uniformly enlarged, the anterior fontanel tense, and the sutures separated. She was admitted to Children's Hospital March 27, 1951. The head measured 42 cm. It transilluminated readily and the red glow could be seen through the pupils, as described by Hamby and associates.[1] There were incoordinate movements of the eyes, the gaze usually being downward. On retinal examination the discs were well defined but pale; the vessels were within normal limits and there were no areas of chorioretinitis.

A tap through the angle of the anterior fontanel revealed clear fluid with 14 cells of which 8 were polymorphonuclear and 6 mononuclear; the protein was 116 mg. per cent; the sugar was 47 mg. per cent. A spinal tap revealed a similar fluid. Neutral methylene blue dye inserted at the time of the ventral tap did not appear in the spinal fluid.

Electroencephalograms were reported upon by Dr. Knox Finley, as follows: "The record consists of a mixed frequency of 5 to 12 per second cycles and a number of slow 1 to 2 per second cycles of moderate amplitude arising from all areas of both hemispheres. Interpretation: The electrocardiogram is probably abnormal for the age of 25 days because of the number of slow 1 to 2 per second waves. The slow waves are diffusely distributed without evidence of focal or lateralized cerebral pathology from the eight scalp electrode placements."

Pneumoencephalograms are of especial interest. The radiologist, Dr. Evelyn Siris, reported in advance the exact conditions revealed at autopsy. Some of the films are reproduced. Fig. 1, taken with the child held head down, indicates that the infratentorial space is small but the tentorium itself is well visualized. Just above the pituitary fossa there is a small soft tissue shadow and in the region of the fourth ventricle there is a faint triangular shadow of decreased density that may represent a moderately dilated fourth ventricle; presumably these shadows represent portions of a midbrain and pons though there is nothing that definitely represents cerebellum. Fig. 2 shows that the falx and the tentorium are present and intact. The opening in the falx where the cerebral hemispheres normally communicate is well visualized and devoid of tissue. There is communication between the right

From Children's Hospital and the Department of Pediatrics, University of California Medical School.

and left halves of the cranial cavity. There is no definite lateral ventricular system and no sizable amount of cerebral tissue. In other views a slender soft tissue density along the anterior inner margin of the left parietal bone may possibly represent a small amount of cerebral hemisphere tissue but it may also represent the somewhat water-laden layers of the meninges.

Cerebral arteriograms were made after injection of Thorotrast into the right common carotid artery by Dr. Mary Mathes. The caliber of the internal carotid artery was about normal. There was immediate reflux into the opposite internal carotid across the anterior communicating arteries, and into the basilar and vertebral arteries on the left across the posterior communicating arteries. The circle of Willis appeared to be intact. The anterior cerebral artery and its branches and the sylvian group of arteries are present but are very thin and elongated (Fig. 3). The sylvian group is displaced far laterad as is seen in extreme hydrocephalus (Fig. 4). The internal carotid itself, including the portion within the cavernous sinus, does not appear unusually narrowed, but immediately thereafter the vessels attain a very narrow caliber, suggestive of atrophy. The vessels of both cerebral hemispheres are visualized since there is ready reflux across the anterior communicating artery. About the orbit and supraorbital area there is a marked fine arteriovenous network. This is drained both by the anterior facial vein and an anomalous cerebral vein which appears too high to be the superior ophthalmic vein but which empties into the cavernous sinus. The ophthalmic arteries and veins appear to be normal. The middle meningeal artery is seen. The external carotid artery is also partly seen.

In the venous phase the superior sagittal sinus, the transverse and sigmoid sinuses, parts of the inferior sagittal, and occipital sinus are easily seen (Fig. 5).

The autopsy was done by Dr. Pearl Smith. It is given in full with the exception of the description of the body. The possible secondary effects, especially upon the endocrine glands, of such marked brain and basal nuclear abnormality warrant a detailed recording.

Thymus: The thymus is large, weighing 17 grams, and is pale and soft. A large area of hemorrhage is present beneath the capsule anteriorly, and there is a subpleural area of hemorrhage that extends to within a few millimeters of the diaphragm.

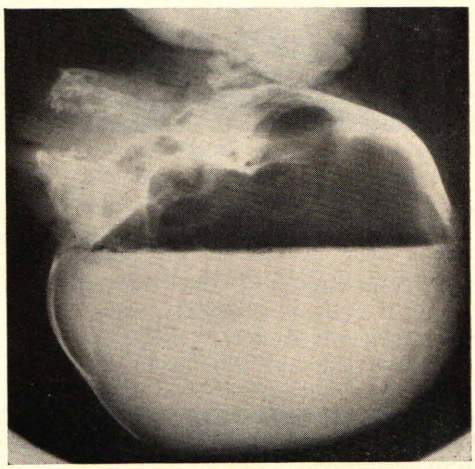

Fig. 1.—Child held head down. Note small infratentorial space, and soft tissue shadow above and behind sella turcica.

Microscopically, it is narrower than usual and numerous phagocytes are distributed through it. The thymocytes are closely spaced. Hassall's corpuscles are numerous and small; their appearance is not unusual.

Pericardium and heart: The pericardial sac contains a normal amount of clear, yellow fluid, and the pericardia are smooth and glistening. The heart, freed from blood, weighs 26 grams. Its proportions are normal and, on section, there are no abnormalities of the valves or septa. The great vessels enter in the normal position. The foramen ovale is normally patent, and there is probe patency of the ductus arteriosus. The

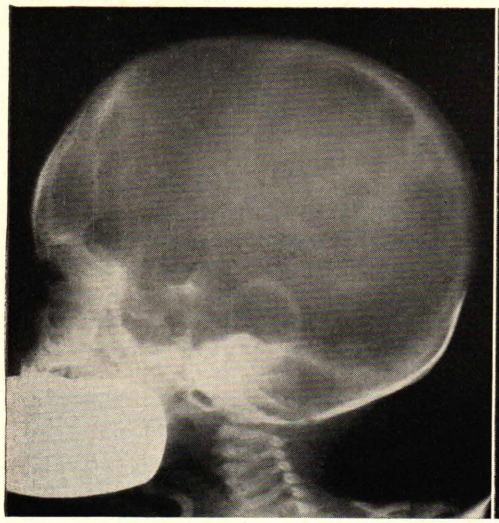

Fig. 2.

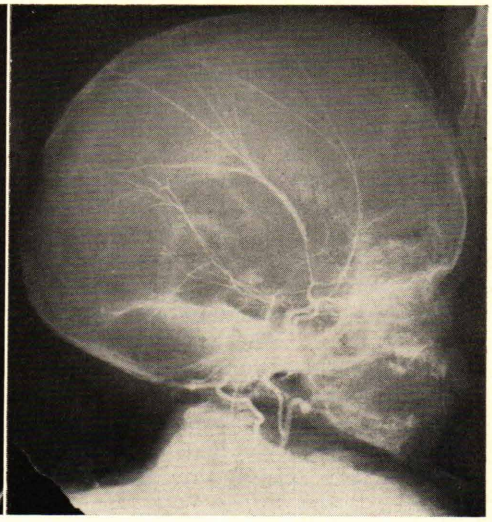

Fig. 3.

Fig. 2.—Lateral projection. Crescentic lower anterior margin of Falx demonstrated. Note tentorium.

Fig. 3.—Arteriogram, lateral. Both internal carotids and vertebral arteries are visualized from a single injection into the right internal carotid artery.

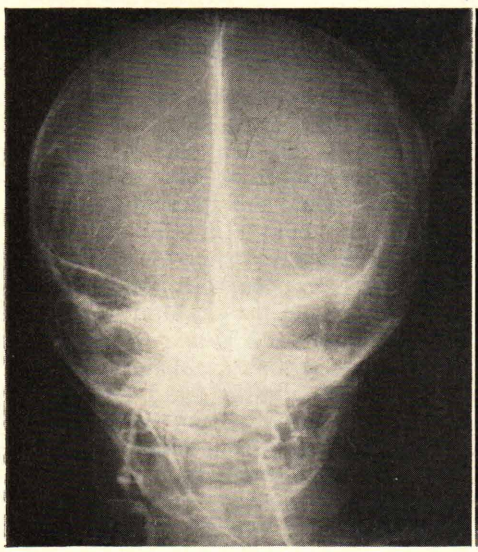

Fig. 4.

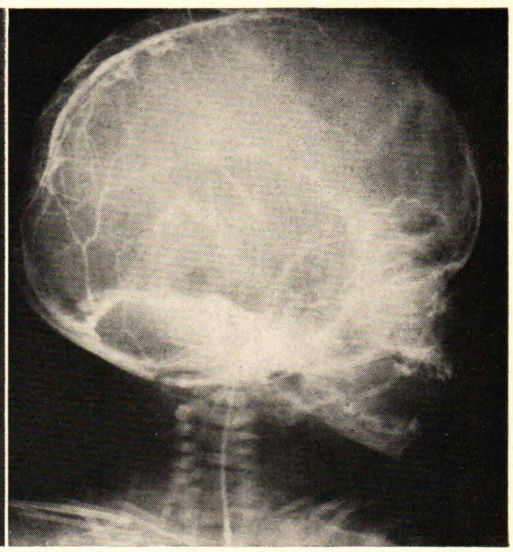

Fig. 5.

Fig. 4.—Arteriogram, anteroposterior. Note lateral displacement of sylvian group.
Fig. 5.—Arteriogram. Dural sinuses are demonstrated.

muscle is firm but pale. Microscopically, the epicardium and endocardium are normal. The cytoplasm of the myocardial fibers is coarsely granular, and the transverse striae are indistinct. The fibrous septa are widened and edematous.

Pleurae and lungs: The pleural cavities are free from fluid. The pleural surfaces of the lungs are smooth and pink. On section, the parenchyma is light red and appears to be normally air containing. Microscopically, in small areas the alveoli are atelectatic but most of them are moderately well expanded.

Peritoneal cavity: The peritoneal cavity contains a normal amount of clear, colorless fluid, and the peritoneal surfaces are smooth and glistening. The organs are in their normal position.

Spleen: The spleen weighs 25 grams. It is soft, smooth, and on section it is light red and the malpighian corpuscles are indistinct. Microscopically, the malpighian corpuscles are moderately enlarged; in a wide zone around them the pulp is filled with red blood cells and numerous large mononuclear phagocytes filled with coarse clumps of brown pigment. The sinuses are narrow and contain very few cells.

Liver: The liver weighs 169 grams and is firm, smooth, and light reddish-tan. On section, the surface is firm, smooth, and uniformly reddish-tan without obvious lobular markings. Microscopically, the portal triads are normal. In the central zone the cells of the liver columns contain small vacuoles and their cytoplasm is coarsely granular; elsewhere the cells are within n o r m a l limits. The Kupffer cells everywhere are filled with coarse brown pigment granules.

Pancreas: The pancreas is normal grossly and microscopically.

Adrenals: The adrenals together weigh 2.5 grams. They are normal in outline and, on section, show the usual cortico-medullary differentiation. Microscopically, the medulla is normal. There is less than the usual amount of differentiation between the fascicular and glomerular zones. The cells of the fascicular zone contain an approximately normal amount of lipoid material. The entire cortex is thinner than usual.

Urinary system: The right kidney weighs 16 grams, the left 17 grams. The capsules strip easily leaving a smooth pale pinkish-tan surface. On section, the architecture of the cortex and medulla appears to be normal, and there are no obvious abnormalities of the pelves, ureters, or bladder. Microscopically, in the subcapsular zone of the cortex an occasional glomerulus is smaller than usual but is still well differentiated. Elsewhere the glomeruli and their tubules are normal.

Gastrointestinal system: There are no apparent lesions of the stomach or intestines grossly or microscopically.

Genitalia are of the normal female infantile type.

Bone marrow: The bone marrow is pale pink. Microscopically, it has a normal cellularity with the usual distribution of cells of various types.

Head: The head is moderately enlarged. The coronal and lambdoid sutures are markedly widened, the coronal to a maximum width of 5.5 cm. and the lambdoid to a maximum width of 3.5 cm.

Brain: The convolutions are completely flattened. In the right cerebral hemisphere the parenchyma in the right frontal region is apparently limited to small soft gray patches. (Fig. 6.) Numerous minute opaque flecks are diffusely distributed through this region. In the parietal and occipital regions the parenchyma is 1 to 2 mm. thick on the right and 2 mm. thick on the left. The posterior surface of the cerebellar hemispheres is irregularly lobulated and cystic and the covering appears to be only arachnoid except at the tips of the temporal lobes which are apparently formed by pia arachnoid only, in an area 4 cm. in diameter on the right and 3 cm. in diameter on the left.

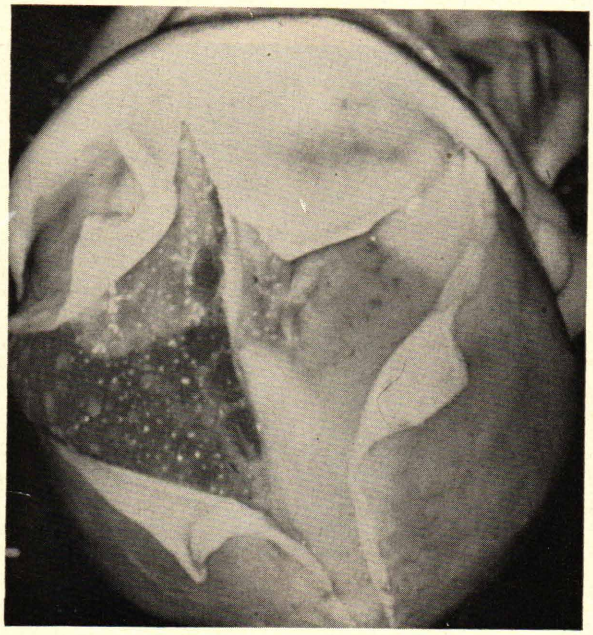

Fig. 6.—Calcified flecks on dura. Pathologist's interpretation was that these were isolated remnants of brain tissue that had calcified. There was no evidence of toxoplasmosis.

Fig. 7.—Photograph of brain post mortem. Note cystic cerebellum.

The basal nuclei are markedly atrophied, with a maximal thickness of 2 mm. at the level of the infundibulum. The ventral part of the cerebellum is 4 cm. wide and 1 cm. thick. There is a marked decrease in size of the pons, medulla, and cord. The pons and medulla are each 1 cm. wide and 1 cm. thick. The cranial nerves are identified but are considerably smaller than usual. The brain weighs approximately 300 grams.

Pituitary: The pituitary gland is normal.

In sections of the cerebral cortex the brain matter has a normal architecture. The decrease in thickness is largely in the white matter and in this region there are numerous areas of edema. In the remaining cerebellum Purkinje's cells are present and the granular cells appear normal. In sections of the basal nuclei there is a marked atrophy of all elements with calcium deposition in the central zone. In the pons and medulla the decrease in size is most marked in the ventral portion due to a marked decrease in the volume of the pyramidal tracts.

Anatomical Diagnosis.—Hydranencephaly, with marked atrophy of the cerebral parenchyma. Hypoplasia of the pons and medulla. Pigmentation of the liver and spleen.

COMMENTS

This study was made with the hope that angiography would give additional information regarding the etiology of hydranencephaly. Authors[2] in general have concluded that to produce the intact cranium and meninges seen in these cases the anatomical structures would have to develop normally until approximately the fifth or sixth month of intrauterine life. It then becomes necessary to find the agent or agents that would dissolve the brain tissue, leaving the cranium, meninges, and other structures intact. The various possibilities listed have been vascular anomalies causing a gradual depletion of the circulation, thrombosis or emboli producing the same effect, infection due to bacteria, viruses or such agents as toxoplasma, toxic agents, particularly bacterial toxins, and congenital deformation of the brain independent of vascular anomalies.

The angiograms reveal an intact circulatory pattern. The vessels are small, the size undoubtedly being the effect and not the cause of the pathology. Incidentally, the network of vessels in the supraorbital area is probably related to the nevus flammeus commonly seen in this area in newborn infants.

The pigment in the spleen and liver must represent the remnants of the destructive process in the brain.

The slight variations in the adrenals and thymus cannot be attributed specifically to the central nervous system changes but may represent bodily changes in adjustment to such pathology.

There is no evidence of inflammatory changes either in the remaining tissues of the central nervous system or in other organs of the body. Tests for toxoplasmosis were negative.

It can be assumed that a sudden catastrophic change in the cerebral tissue resulting in this amount of destruction late in the fetal development would cause death.

Hydranencephaly and porencephaly are pathological states that may be produced by any of the mentioned causes and agents. In the patient here reported it would seem that the most likely etiology is on a congenital basis. Multiple cyst formation could be identified in the cerebellum. It is plausible that similar small, closed,

secreting cavities or cysts appearing early in the development of the brain might proceed to increase in size, coalesce, and finally result in complete destruction of the brain in what amounts to universal porencephaly.

Attention is called to the simple device of taking the picture of a pneumoencephalogram in the upside down position. A relatively small amount of air reveals the extent of the pathology.

SUMMARY

A case of hydranencephaly in which angiography was done is reported to show that in this patient the vascular pattern like that of the cranium and meninges is intact.

The picture of the pneumoencephalogram taken with the patient upside down reveals clearly the extent of the destruction.

There was no evidence of vascular anomaly, of infection, or toxic inflammation to indicate these factors as a role in etiology, thus leaving congenital cystic formation within the cerebral tissue as the most likely cause.

REFERENCES

1. Hamby, W. B., Krauss, Ruth F., and Berwick, W. F.: Hydranencephaly: Clinical Diagnosis, Pediatrics 6: 371, 1950.
2. Watson, Ernest H.: Hydranencephaly, Am. J. Dis. Child. 67: 282, 1944.

CEREBRAL ANGIOMATOSIS

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CEREBRAL ANGIOMATOSIS

Report of a Case in an Infant, With a Review of the Literature

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THE diagnosis of cerebral angiomatosis in a 7-week-old infant prompted a review of the literature in an attempt to delineate better the symptoms and signs that should alert the physician to the possibility of arteriovenous malformations in the child. A fair proportion of these abnormalities are amenable to surgery and, when they are, surgery should be performed before irreparable damage has taken place.

REVIEW OF LITERATURE

The first presumptive clinical diagnosis of a cerebral angioma was made in 1898 in a patient with left spastic hemiparesis and a bruit, audible over the entire head. Since the introduction of angiography it has become evident that intracranial angiomas or, more correctly, arteriovenous malformations are not rare.

According to Paterson,[1] most intracranial angiomas are to be found on the lateral aspects of the cerebral hemispheres and in particular near the Sylvian fissure. Less commonly they are situated on the medial aspects of the hemispheres, in the corpus callosum, the orbits, within the ventricles, in the posterior fossa, or near the midline in basal structures. Noran[2] gave a pathological classification of the intracranial vascular tumors and malformations closely correlated with the types of vascular structure, thereby increasing the understanding of the relationship between their clinical and their pathological aspects. Dandy[3] was one of the first to describe cerebral arteriovenous angiomas. In his 8 cases, focal epileptic attacks were present and a gradually progressive motor or sensory hemiplegia of the affected side developed. Cerebral hemorrhage, the principal cause of death, occurred in about 40 per cent.

In the same year, Cushing, and Baily and Dandy,[3, 4] reported 22 cases of which 17 were arterial angiomas with arteriovenous connections and 5 were carotid cavernous fistulas. The most common symptom was headache. Epileptiform seizures were reported in 6 patients, nausea and vomiting was observed in 8, bruit in 5, and intracranial calcification in 4. The youngest patient was a 6-year-old girl with convulsions, mental retardation, and right-sided paresis and ataxia.

In 1936 another monograph appeared, in one section of which Toennis[5] considered a number of patients with arteriovenous aneurysms. He recorded a series of 22 cases in some of which a diagnosis by arteriography

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was made before operation. An audible bruit was found in 4 cases and several had Jacksonian attacks.

In 1937, Jaeger, Forbes, and Dandy[6] described the occurrence of a bilateral congenital cerebral arteriovenous communicating aneurysm diagnosed at autopsy in a 3-year-old child.

In 1948, Olivecrona and Riives[7] reported 42 cases of arteriovenous aneurysm. The cardinal signs were epileptic fits, subarachnoid hemorrhage, and hemiplegia. A systolic bruit was heard in 8 cases with characteristic calcifications. The first symptoms usually occurred during the second or third decade.

In 1953, MacKenzie[8] presented 50 cases where the presence of a cerebral angioma had been proved by arteriography. The diagnosis was made in this series between the ages of 12 and 59 years. Analysis of the presenting complaint showed this to be epilepsy in 16 cases, hemorrhage in 15, headache in 12, hemiparesis in 6, and a pulsating exophthalmus in one. One patient with a left-sided angioma had a right-sided infantile hemiplegia. According to the studies of MacKenzie,[9] only 10 per cent of the patients had a relevant symptom in the first decade of life, in 70 per cent before the age of 30. About 90 per cent were discovered before the age of 40. If there has been no recent rupture, the spinal fluid is normal. X-rays of the skull and electroencephalogram are often normal.

In 1953, Toennis and Lange-Cosack[10] also described 76 patients with arteriovenous angiomas of the brain. In 55 patients chronic psychopathic syndromes were seen. In 35 cases mental deterioration occurred. All patients with frontal, temporal, and diffuse angiomas as well as a high percentage of the centroparietal angiomas had lasting psychic changes. There also were frequent epileptic attacks. The treatment of choice was total extirpation before cerebral atrophy had developed. Ligation of the carotid artery was discarded as an inadequate procedure.

In 1956, Walker[11] studied a series of 84 cases of intracranial vascular malformations verified by angiography in 9, operation in 76, and autopsy in 17. Approximately 80 per cent of the patients had a history of symptoms for more than two years. Epilepsy was the presenting complaint in 40 of the 64 patients with supratentorial lesions. In 26 cases the seizure was of the Jacksonian type. The next most common symptom was a neurologic deficit—in 13 cases a hemiparesis—usually of the spastic type. Subarachnoid hemorrhage was manifest in 14 cases. Intracranial hypertension occurred in 2 patients associated with cranial nerve palsies, notably a sixth nerve paralysis. Noises in the head were noted by 8 patients. In 5 of these cases a bruit could be heard on auscultation. Hemorrhage was reported in 20 per cent. Other authors reported hemorrhage as the first symptom in a high proportion of patients with intracranial vascular anomalies, e.g., 56 per cent in the series of 24 cases of Gould, Peyton, and French,[12] and 54 per cent in the presentation of 37 cases by Anderson and Korbin.[13]

Crawford and Russell[14] discussed 20 cases of spontaneous cerebral and cerebellar hemorrhage in 15 children and adolescents and 5 adults due to small arteriovenous or venous hamartomas of the brain. Clinically the

cases presented either as catastrophic apoplexies or, less acutely, simulating rapidly growing neoplasms.

Potter[15] reported 58 cases of cerebral angiomas verified either at operation, by angiography, or at autopsy. In his series, only 22 per cent presented a cranial bruit and in only two were there extracranial vascularities. Hemorrhage occurred in 82 per cent and was the presenting symptom in 58 per cent.

In a study of 148 patients with spontaneous subarachnoid hemorrhage, Falconer[16] pointed out that arteriovenous malformations are the second most common cause of subarachnoid hemorrhage, accounting for about 5 to 10 per cent. More malformations manifest themselves by epilepsy than by hemorrhage. The association of epilepsy, the occurrence of bleeding in childhood, or a history of attacks of bleeding extending over many years is more suggestive of malformation than of aneurysm. All his 12 patients who underwent operation survived and were benefited by the surgery.

A series of 110 patients with intracranial angiomas have been seen by McKissock[17, 18] at the National Hospital and St. George's Hospital, London. The cardinal symptoms were those due to a subarachnoid or intracerebral hemorrhage (42 per cent), with or without epilepsy and periodic migrainous headaches. The youngest patient to develop epilepsy in this series was a 1-year-old child. In some cases a progressive hemiparesis with insidious onset was seen. Intracranial bruit was present in 38 per cent, focal neurological signs in 69 per cent, while papilledema and dementia were not common. Associated vascular anomalies and complications were cirsoid aneurysms of the scalp, intracranial aneurysms, and chronic subdural hematomas. Two patients of this series had arteriovenous malformations involving the orbit with increasing proptosis of the affected eye and a pulsating bruit referred to the ear on the same side. Total excision was reported as the ideal form of treatment. In 35 patients, McKissock was able to remove the malformation completely. Ligation of cortical feeding vessels was of value in certain cases but ligation of the common or internal carotid artery did not appear to be of value in treatment. Also the results of radiotherapy were disappointing.

The frequency of a bruit varied in reported series from 15 per cent[12] to 50 per cent,[8] averaging 22 per cent. A systolic bruit is audible in a certain proportion of normal infants and young children, also in some patients with small saccular aneurysms,[19] over certain vascular tumors, in carotid-cavernous fistula,[20, 21] and contralateral to thrombosed cervical carotid artery.[22] Hamburger[23] stated that cephalic murmurs in children are prevalent particularly during the second year of life when they occur two and a half times as frequently as in the first year of life. Ford[24] attributed these benign murmurs to anemia. The youngest case in the series of Gould, Peyton, and French[12] was diagnosed at the age of 6 weeks. The parents had observed a rapid growth of the head. The arteriovenous malformation was in the left cerebellar hemisphere where it had produced a block of the flow of the cerebrospinal fluid.

Logue and Monckton[25] reported 9 patients with angiomas in the posterior fossa. Two were located in the cerebellum with cerebellar apoplexy, sixth nerve involvement and scattered pyramidal tract signs. The remaining seven were located in and about the brain stem. Of these 7 patients, 4 had subarachnoid hemorrhage, 2 progressive neurological signs, and 1 acute obstructive hydrocephalus. The general neurological picture was that of a progressive disorder of the third to the eleventh cranial nerves, symptoms of pyramidal tract involvement or cerebellar ataxia and tremor. Three patients had an intracranial bruit.

Rosner[26] reported 34 cases of cerebral palsy of which 33 were due to vascular anomalies. In these 33 children Rosner found cerebral venous angioma at the lower angle of the Sylvian fissure on one or both sides. Craniotomy was performed and the angiomatous vessels were cauterized with the electrocautery. The 29 children who survived the operation showed improvement of spasticity and athetosis as well as of epilepsy and of drooling.

A case of bilateral optic nerve atrophy and blindness of the left eye without any other signs or symptoms was presented by Band.[27] The cause was a cerebral arteriovenous aneurysm.

Laur[28] reported a giant cerebral angioma with anomalies of the great arteries and multiple saccular aneurysms in a 15-year-old boy who developed epileptic seizures at the age of 15. He had a right pulsating exophthalmus which had been present since birth; he also had an internal hydrocephalus, debility, and cardiac enlargement. Cerebral angiography revealed an arteriovenous aneurysm of the left hemisphere measuring 11.5 by 8 by 7 cm. and also saccular arterial aneurysms on the right side, together with anomalies of the circle of Willis and multiple aneurysms of the right internal carotid.

Groethuysen, Bickford, and Svien[29] gave a detailed analysis of the electroencephalogram in 18 patients with arteriovenous anomalies of the brain. They as well as Gillingham[30] and others reported that the diagnostic value of the electroencephalogram in evaluating suspected cases is limited. Low-voltage bilateral abnormalities in theta or delta range with one-sided predominance were found in the majority of arteriovenous anomalies. In view of the high localizing value of ictal and postictal focal electroencephalographic changes, the more frequent use of activation methods to induce a seizure was suggested.

Ferey[31] reported 10 cases of central angioma. In 5 patients the initial symptom was progressive hemiparesis; in 4 a complicating subarachnoid hemorrhage was accompanied by the rapid onset of focal signs, usually a hemiparesis. One patient only had epileptic seizures. This boy started to have monthly grand mal seizures at the age of 2 years. Nine patients had contralateral hemiparesis accompanied in 5 instances by sensory defects and in 4 by a partial or complete homonymous hemianopsia, by pupillary abnormalities in 3, ptosis and external ocular palsies in 4, and nystagmus in 2 cases. In only 2 of the 10 patients was an intracranial bruit heard. In 8 of the 10 cases the routine skull film was reported to be

normal. In 9 of the 10 patients the angioma was diagnosed by angiography and was thought to lie in the neighborhood of the thalamus, basal ganglia, and internal capsule. In one the diagnosis was made by post-mortem examination.

Rosenberg[32] presented a 4-month-old infant with alternating Jacksonian seizures, opisthotonus, and progressive hydrocephalus. Angiography revealed an arteriovenous malformation with communication between the left anterior cerebral artery and the great vein of Galen. Autopsy showed an angiomatous malformation on the medial aspect of the left cerebral hemisphere.

Arteriovenous malformations in the region of the pineal gland are rare. The first reported case of arteriovenous malformation drained by the vein of Galen was described by Jaeger, Forbes, and Dandy,[6] and reports of Alpers,[33] Oscherwitz,[34] Boldrey,[35] Olivecrona,[7] Gillingham,[30] and French[36] followed. French and Peyton[36] reported 5 cases of arteriovenous malformation in the region of the pineal gland. Although these are unfavorable for removal because of their location, nevertheless they are operable when it is possible to demonstrate their arterial supply by angiographic studies. The only signs may be increased intracranial pressure with slowly developing hydrocephalus.

In 1949, Norlen[37] reported 10 patients with arteriovenous anomalies of the brain in whom radical removal resulted in restoration to normal in 7, and in improvement of neurologic defects in 3. In one patient mental function was reported to be improved some months after operation.

Bassett[38] presented 18 cases of cerebral arteriovenous anomalies. He recommends direct surgical attack and does not believe that irradiation has a place in treatment.

Pilcher, Meacham, and Cobb[39] reported 11 cases of intracranial angiomatous malformations and recommended surgical removal if the lesion is accessible. After occlusion of the arterial radicals, the venous tributaries are clipped and the lesion removed. Lobectomy as an alternative plan of removal may be used when the angioma is located in the frontal or occipital lobe.

The works of Olivecrona and Riives,[7] of Norlen,[37] of McKissock,[40] of Jaeger,[41] and of Toennis and Lange-Cossack[10] indicate that the treatment of choice is now total excision of the malformation. Where this is not practicable, as in deep-seated lesions, the feeding artery can be occluded and any associated intracerebral clot evacuated.

CASE REPORT

A 7-week-old white female infant entered the hospital on June 25, 1957, because of a progressive swelling of the right eye. The child was born after a full-term pregnancy that was normal except for a mild kidney infection. Delivery was with low forceps. Birth weight was 8 pounds 11 ounces. There was no cyanosis or respiratory difficulty. Immediately after delivery the mother noted a swelling of the right eye. This was thought to be due to the forceps. However, the swelling increased and the eye deviated to the right and appeared prominent. On the day of admission the infant had twitchings of her face, mouth, and arms. These twitchings lasted up to ten minutes and had occurred about six times prior to admission. The child held

her head to the right side and seemed more sleepy than usual. There had been no vomiting.

Family history was not contributory. The four siblings aged, respectively, 10, 8, and 3 years, and 16 months were well.

Physical examination revealed a 7-week-old infant with an obvious deformity of the right eye. The head was held toward the right. Head circumference was 40.5 cm. The anterior fontanel was bulging and measured 5 by 7 cm. The posterior fontanel was also bulging and measured 4 by 5 cm. The veins on both sides of the head, but particularly over the right hemicranium, were engorged. There was a small, soft swelling above the superior portion of the sagittal suture extending to the right. In addition, a soft discrete swelling on the right side of the forehead was noted and a diffuse soft swelling involving the bridge of the nose, the right supraorbital ridge, and the lateral portion of the right eye. A loud to-and-fro bruit was heard over the right side of the skull. This bruit could be obliterated by compression of the right carotid. The right eye was proptosed and showed chemosis of the conjunctiva. Both pupils were deviated to the right. The right pupil was irregular and larger than the left and reacted sluggishly to light. Fundoscopic examination revealed dilated veins; however, the margins of the discs were well defined and there was no evidence of papilledema. Examination of the chest was negative. The liver was palpable 2 cm. below the right costal margin. The deep tendon reflexes were normal, and no stiffness of the legs, back, or neck was noted. There was no limitation of motion. Temperature was 38.4° C., pulse 136, respiration 70, weight 8 pounds 14 ounces.

Examination of the blood revealed a hemoglobin of 6.9 Gm. per 100 ml., red blood cells 2,240,000 and a white cell count of 8,700. The hemoglobin was raised to 13.8 Gm. by a blood transfusion (135 c.c.). The urine was normal. Serum bilirubin was normal. Kahn and Kolmer tests were negative. Cerebrospinal fluid was slightly xanthochromic, with 81 red blood cells, and 24 white blood cells with 20 neutrophils and 4 lymphocytes, protein was 86 mg. per cent, glucose 44 mg. per cent. There were no tumor cells or organisms seen. A ventricular tap 8 weeks later revealed 214 mg. per cent protein. On subdural taps, bright red blood was obtained with icteric supernatant fluid. The smear showed crenated red blood cells. No organisms were seen on stained smear and the culture was negative. Biopsy of the tumor at the right side of the scalp was performed by Dr. Pieter A. de Vries. It showed a hemangioma.

Skull films and pneumoencephalograms revealed increased intracranial pressure. There was no definite evidence of internal hydrocephalus. However, external hydrocephalus appeared to be present and a communicating hydrocephalus was suspected. A small area of bone destruction in the right parietal bone was seen. A subsequent ventriculogram revealed an internal hydrocephalus. A cerebral angiogram (Fig. 1) disclosed an arterial venous malformation which shunted the blood received from the internal carotid artery away from the right cerebral hemisphere. There appeared to be two parts to this malformation. One was identified as a malformation in the region of the cavernous and circular sinus and the other apparently connected through the great vein of Galen and the straight sinus to the right side of the posterior fossa subtentorially. The malformation was extensive and deep. The tentorial and infratentorial dural sinuses were greatly enlarged. The size of the lesion precluded operation.

On the day after admission the child had another convulsion with tonic-clonic movements of the left arm and leg both held in partial flexion. The seizure lasted about two

minutes. Subsequently the infant had repeated episodes of generalized twitchings, starting on the right with convulsions of the right arm and leg. Later a left hemiparesis was noted. weeks after admission. Extreme opisthotonus and high-pitched cry were noted. The infant died at the age of 4½ months, 12 weeks after admission.

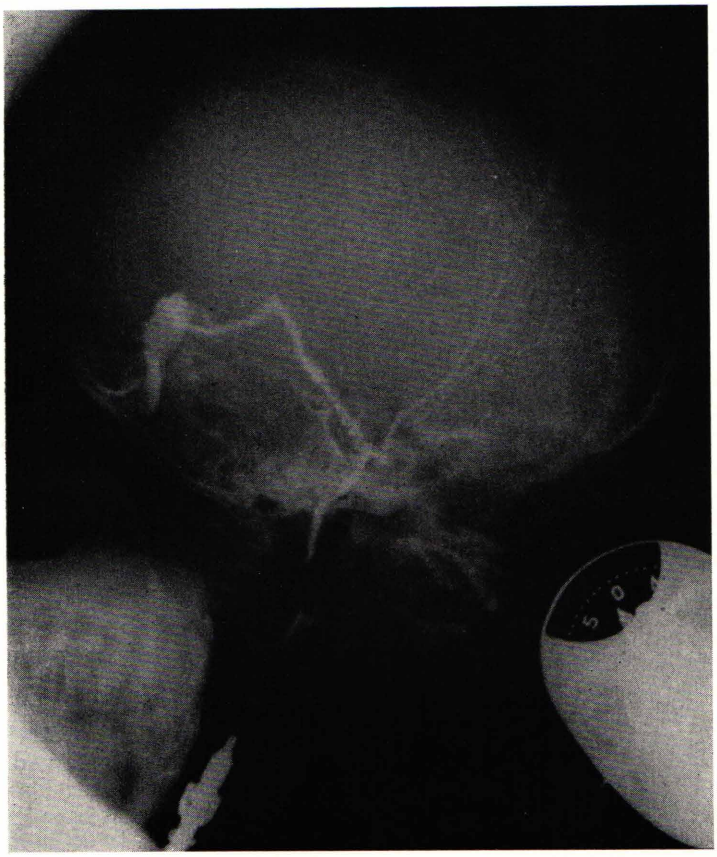

Fig. 1.

The head was constantly turned to the right side. Fundoscopic examination revealed a progressive right optic nerve atrophy. The course was steadily downward, with fever of 38.5° up to 39.2° C. without obvious signs of infection. However, during the last ten days before exitus the temperature was subnormal, even below 34° C. Pulse rate varied between 140 and 230, respirations between 40 to 96. The head circumference increased rapidly from 40.5 cm. on admission to 44.5 cm. at four weeks, 47 cm. at eight weeks, and 48 cm. at 12

Post-mortem examination was done by Dr. Samuel T. Nerenberg and the detailed studies of the brain were made by Dr. Knox Finley.* Only the brain will be described, as the rest revealed no pertinent pathology. The left cerebral hemisphere appeared about 20 to 25 per cent larger and somewhat paler than the right. The grayer right and smaller cerebral hemisphere had two large vessels extending from the approximate region

*Graduate Training Grant in Neuropathology (Stanford), National Institute of Health. Division Neurological Diseases and Blindness.

of the falx down along what would appear to be the central fissure. It terminated in an ill-defined dark grayish area lying on the lateral portion of the frontal lobe, measuring approximately 3 cm. in diameter. The surface of this grayish area, which was firmer than the surrounding brain parenchyma, was striated by numerous tortuous vessels. The posterior two-thirds of the falx were much thicker and firmer than normal. Posteriorly to the region of the vein of Galen and at the point where the falx joins the tentorium, there was considerable thickening by a combination of a firm and fibrinous tissue between the two outer layers of the falx and the tentorium, becoming eventually a tumor mass greatly distending the terminal portion of the falx but primarily the tentorium. This tumor mass measured approximately 10 cm. in an anterior-posterior-inferior direction and 3 to 4.5 cm. in width. Running through or near the center of the tumor mass was a large channel, rather tortuous in its course, the lumen measuring 5 to 8 mm. in diameter. The brain stem was not unusual in the gross though there had been some distortion from the terminal hemorrhage of the above-described tumor and possibly in fixation. However, the subarachnoid spaces at the base of the brain stem, cerebellum, and, for that matter, part of the base of the cerebral hemispheres had a yellowish gold-brown discoloration. The right cerebellar hemisphere was slightly more than half the size of the left.

It was not possible from the very friable, distended, collapsed brain to find a vessel which communicated with the internal carotid or main cerebral vessels to the tumor mass in the posterior tentorium and falx. The large vessels over the central portion of the right cerebral hemisphere which had now been separated from its apparent connection with the tentorium did appear to have penetrated the falx about 1 to 2 cm. above and anterior to where the falx became continuous with the tentorium. On opening the temporal and occipital horn of the right ventricle there was a very rich and tortuous mass of subependymal vessels seemingly involving most of the subependymal region of these portions of the right lateral ventricle. On opening the anterior horn of the right lateral ventricle, the anomalous vascular structures could likewise be traced into this portion of the ventricle. These vessels were continuous with the 3 by 4 cm. grayish mass, with tortuous vessels described from surface observations of the lateral part of the right frontal lobe. Opening the ventricle of the left hemisphere showed a tremendous dilatation throughout, the overlying white matter and cortex measuring in thickness not more than 0.5 cm. plus-minus.

An attempt was made to find some direct channel between the circle of Willis and the proximal portions of the main cerebral vessels, that is, the anterior middle and posterior cerebral vessels, but, after fixation, no direct communication could be found. However, it should be pointed out that because of the great difficulties in removing such a brain with tremendous internal hydrocephalus, such communication could have been separated. Frontal sections through the brain stem and cerebellum revealed a tremendously dilated fourth ventricle. Sections of the spinal cord were normal at cervical, thoracic and lumbosacral levels.

Numerous sections were examined with the Nissl and hematoxylin and eosin stains. Sections from the falx show a conglomeration of vessels, most of which were of thick-walled (arterial) type. A number of the vessels were thrombosed. Hemosiderin pigment and mononuclear leukocytes were present throughout the sections. Many of the vessels contained calcification in their walls. Sections through the cortex showed the pia-arachnoid meninges to contain numerous anastomosing and calcified

thin-walled vessels; many of the lumina were thrombosed. The underlying cortex contained scattered small hemorrhages and collections of hemosiderin pigment. In a number of areas there was proliferation of the astrocytes and microglia. Sections through the cerebellum and pons showed marked deformity and flattening. Much of the cerebellar tissue was replaced by a vascular malformation made up of numerous, dilated, blood-filled and thin-walled spaces separated by parenchyma. Scattered gliosis and occasional hemorrhages were seen between the vessels. The cerebellum and cortex showed degenerative changes. Sections through the medulla showed collections of hemosiderin pigment on the surface. Sections through the spinal cord (cervical portion) showed a collection of mononuclear cells and hemosiderin pigment surrounding the meninges. Sections through the thoracic cord showed subdural hemorrhages, degenerating leukocytes and hemosiderin.

Diagnosis.—Extensive vascular malformation of the central nervous system (angiomatosis of meninges and brain). Subarachnoid and subdural hemorrhage recent and old.

DISCUSSION

Although cerebral vascular anomalies are congenital in origin, manifestations of their presence are seldom evident at birth. In this review, only one other was reported as young as our patient. One author notes that less than 10 per cent manifest themselves in the first decade. One child had convulsions starting at 4 months, another showed symptoms at 1 year, one was diagnosed at 2 years, one at 3 years, and another at 6 years. Curiously there is very little mention of associated visible clues. In our case the biopsy of the soft tumor mass virtually made the diagnosis. It was then confirmed and delineated by angiography and ventriculograms.

A pulsating exophthalmus was noted in one case and a cirsoid aneurysm of the scalp in another. In the majority of cases, diagnosis must be made on symptomatology and by angiography.

The presenting symptoms as gleaned from the review of the literature may be grouped as follows:

1. Palsies are often unilateral, the patient presenting with a hemiplegia which may involve both the motor and sensory areas and often shows progressive signs. Palsies of the cranial nerves have been noted, notably of the third, sixth, and eleventh. Optic atrophy occurs at times.

2. Convulsions or epileptic seizures are commonly recorded as a first sign of nervous system pathology. The attacks may be the Jacksonian type, or generalized. They may occur at long intervals or recur frequently. In some instances they date back to childhood.

3. Cerebral or subarachnoid hemorrhage at any time in life may be due to angiomas.

4. Mental deterioration is frequently noted and is usually progressive.

5. Psychopathic changes are not uncommon.

6. Progressive hydrocephalus occurs when spinal fluid flow is obstructed.

7. Subjective symptoms may be those associated with increased pressure, such as headache and vomiting. In addition, noises in the head may be a complaint.

8. Objective findings have been optic atrophy, nystagmus, bruits over

the head, exophthalmus, calcifications shown by plain x-ray, and occasionally outward signs such as hemangiomata or abnormal vascularity. The anomaly not infrequently is amenable to surgery. Extirpation of the mass may be possible; sometimes hemispherectomy is indicated. Occasionally ligation of vessels leading to the mass may give some relief. Ligation of the carotid artery is inadequate treatment. Radiation is of no value.

SUMMARY

The clinical history and autopsy findings are presented in a child with cerebral angiomatosis. The pertinent literature has been reviewed. General symptoms and signs of such lesions are paresis, epilepsy or convulsions, hemorrhage, less commonly chronic headache, mental deterioration, cranial bruits, visual field defects, diplopia due to sixth nerve paralysis, and symptoms secondary to increased intracranial pressure.

Final diagnosis usually can be made by intracranial angiography. The treatment of choice is radical surgical excision or obliteration and should be carried out whenever feasible. The value of intracranial arterial ligation or incomplete removal of the anomaly may be questioned. Radiation is of no value.

REFERENCES

1. Paterson, J. H.: Central Angiomas, Schweiz. Arch. Neurol. u Psychiat. **75**: 392, 1955.
2. Noran, H. H.: Intracranial Vascular Tumors and Malformations, Arch. Path. **39**: 393, 1945.
3. Dandy, W. E.: Cerebral Arteriovenous Angiomas, Arch. Surg. **17**: 190, 1928.
4. Cushing, H., and Baily, P.: Tumors Arising from the Blood Vessels of the Brain: Angiomatous Malformations and Hemangioblastomas, Springfield, Ill., 1928, Charles C Thomas.
5. Toennis, W.: In Bergstrand, H., Olivecrona, H., and Toennis, W.: Gefaessmissbildungen und Gefaessgeschwuelste des Gehirns, Leipzig, 1936, Georg Thieme.
6. Jaeger, J. R., Forbes, R. P., and Dandy, W. E.: Bilateral Congenital Cerebral Arteriovenous Communication Aneurysm, Tr. Am. Neurol. A. **63**: 173, 1937.
7. Olivecrona, H., and Riives, J.: Arteriovenous Aneurysms of the Brain. Their Diagnosis and Treatment, Arch. Neurol. & Psychiat. **59**: 567, 1948.
8. MacKenzie, I.: The Clinical Presentation of the Cerebral Angioma, Brain **76**: 184, 1953.
9. MacKenzie, C. K.: The Diagnosis of Cerebral Angioma, Postgrad. M. J. **30**: 631, 1954.
10. Toennis, W., and Lange-Cosack, H.: Arteriovenous Angioma of the Brain. Deutsche Ztschr. Nervenh. **170**: 460, 1953; **171**: 416, 1954.
11. Walker, A. E.: Clinical Localization of Intracranial Aneurysms and Vascular Anomalies, Neurology **6**: 79, 1956.
12. Gould, P. L., Peyton, W. T., and French, L. A.: Arteriovenous Malformations of the Brain, Bull. Univ. Minn. Hosp. **26**: 611, 1955.
13. Anderson, F. M., and Korbin, M. A.: Arteriovenous Anomalies of the Brain. A Review and Presentation of 37 Cases, Neurology **8**: 89, 1958.
14. Crawford, J. V., and Russell, D. S.: Cryptic Arteriovenous and Venous Hamartomas of the Brain, J. Neurol., Neurosurg. & Psychiat. **19**: 1, 1956.
15. Potter, J. M.: Angiomatous Malformations of the Brain: Their Nature and Prognosis, Ann. Roy. Coll. Surgeons England **16**: 227, 1955.
16. Falconer, M. A.: Surgical Pathology of Intracranial Hemorrhage Due to Aneurysms and Arteriovenous Malformations, Proc. Roy. Soc. Med. **47**: 693, 1954.
17. McKissock, W.: Infantile Hemiplegia, Proc. Roy. Soc. Med. **46**: 431, 1953; and Zentralbl. Neurochir. **14**: 42, 1954.
18. Paterson, J. H., and McKissock, W.: A Clinical Survey of Intracranial Angiomas With Special Reference to Their Mode of Progression and Surgical Treatment, Brain **79**: 233, 1956.
19. Richardson, C., and Kofman, O.: Cranial Bruit With Intracranial Saccular Aneurysms, Tr. Am. Neurol. A. **76**: 151, 1951.
20. Osler, W.: On the Systolic Brain Murmur of Children, Boston Med. & Surg. J. **103**: 29, 1880.
21. Dodge, H. W.: Cephalic Bruits in Children, J. Neurosurg. **13**: 527, 1956.
22. Fisher, C. M.: Cranial Bruit Associated With Occlusion of the Internal Carotid Artery, Neurology **7**: 299, 1957.
23. Hamburger, L-P.: Head Murmurs, Am. J. M. Sc. n.s. **181**: 756, 1931.

24. Ford, F. R.: Diseases of Nervous System in Infancy, Childhood and Adolescence, ed. 2, Springfield, Ill., 1944, 1086, Charles C Thomas.
25. Logue, V., and Monckton, G.: Posterior Fossa Angiomas. A Clinical Presentation of Nine Cases, Brain **77**: 252, 1954.
26. Rosner, S.: Venous Angioma as a Cause of Cerebral Birth Palsy, J. Nerv. & Ment. Dis. **122**: 276, 1955; and J. Internat. Coll. Surgeons **22**: 94, 1954.
27. Band, R. I.: Optic Atrophy Caused by an Arteriovenous Angioma, A. M. A. Arch. Neurol. & Psychiat. **67**: 655, 1952.
28. Laur, A.: Cerebrales Riesenangiom mit Anomalie der grossen Arterien und multiplen sackförmigen Aneurysmen, Deutsches. Arch. klin. Med. **200**: 236, 1953.
29. Groethuysen, U. C., Bickford, R. B., Svien, H. J.: The Electroencephalogram in Arteriovenous Anomalies of the Brain, A. M. A. Arch. Neurol. & Psychiat. **74**: 506, 1955.
30. Gillingham, J.: Arteriovenous Malformations of the Head, Edinburgh M. J. **60**: 305, 1953.
31. Ferey, D.: L'Hémisphèrectomie cérébrale dans le traitement de l'hémiplégie et de certaines encéphalopathies infantiles. Réflexions à propos de deux cas personnels opérés, Presse méd. **64**: 81, 1956.
32. Rosenberg, A. W.: A Vascular Malformation Drained by the Great Vein of Galen, Bull. Los Angeles Neurol. Soc. **20**: 196, 1955.
33. Alpers, B. J., and Forster, F. M.: Arteriovenous Aneurysm of Great Cerebral Vein and Arteries of Circle of Willis, Arch. Neurol. & Psychiat. **54**: 181, 1945.
34. Oscherwitz, D., and Davidoff, L. M.: Midline Calcified Intracranial Aneurysm Between Occipital Lobes, J. Neurosurg. **4**: 539, 1947.
35. Boldrey, E., and Miller, E. R.: Arteriovenous Fistula (Aneurysm) of the Great Cerebral Vein (of Galen) in the Circle of Willis. Report on 2 Patients Treated by Ligation, Arch. Neurol. & Psychiat. **62**: 778, 1949.
36. French, L. A., and Peyton, W. T.: Vascular Malformations in Region of the Great Vein of Galen, J. Neurosurg. **11**: 488, 1954.
37. Norlen, G.: Arteriovenous Anomalies of the Brain. Report of Ten Cases of Total Removal of the Lesion, J. Neurosurg. **6**: 475, 1949.
38. Bassett, R. C.: Surgical Experiences With Arteriovenous Anomalies of the Brain, J. Neurosurg. **8**: 59, 1951.
39. Pilcher, C., Meacham, W. F., and Cobb, C. A.: The Surgical Treatment of Intracranial Angiomatous Malformations, Surgery **28**: 342, 1950.
40. McKissock, W.: Intracranial Angiomata, Ann. Roy. Coll. Surgeons England **7**: 472, 1950.
41. Jaeger, R.: Arteriovenous Aneurysm of the Brain Cured by Ligation of the Left Middle Cerebral Artery: Report of a Case, Arch. Neurol. & Psychiat. **64**: 745, 1950.

ABNORMALLY SMALL HEAD SIZE AND INTELLECT IN CHILDREN

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Abnormally small head size and intellect in children

Measurements of 212 microcephalic children show them to be consistently retarded in height when compared by age and sex with 12,000 healthy children in the public schools. Abnormally small head sizes in microcephalic children have a significant correlation with intelligence quotients when degrees of microcephaly are plotted against degrees of mental retardation as measured in intelligence quotients.

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STUDIES of normal head circumference for North American infants have been published by Silver and Deamer[1]; Nellhaus[2] reviewed the world literature since 1948 for Negro, Japanese, Alaskan, Eskimo, and Russian children; and Pryor[3] reported the findings in 12,000 American-born Caucasian children. Many other studies of normal children have been reported.

In his books on the dolphin, Lilly[4, 5] discusses the relationship between brain size and intellect. He used acquisition of speech as a measure of intellect and cited McCarthy's[6] study to show that a positive relationship exists between age, brain weight, and speech.

The size of the human head is determined by the growth of the brain except in cases of hydrocephalus, craniosynostosis, or other cranial pathology. Failure of the head to grow has been linked with mental retardation by a number of authors including O'Connell and associates,[7] and Book and associates.[8] Severe brain infections such as encephalitis, meningitis, or toxoplasmosis early in life may prevent further growth of the head, as reported by Thelander and Goebel.[9]

Anthropometric measurements of 181 children with Down's syndrome, reported by us,[10] revealed consistently small head circumferences at every age when compared with normal children. Their heads ceased to grow after the age of approximately 3 years, and they were short in stature.

Bailit and Whelan[11] reported an anthropometric study of body size correlated with intelligence quotients in retarded children. Compared to normal controls, the retarded children were consistently smaller.

Mosier and associates[12] studied 2,027 mentally defective patients in an institution and concluded that, for 4 main classifications, linear growth was stunted to a degree

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Table I. Age distribution to nearest birthday and ranges of head circumferences and I. Q.'s.

Age (years)	*No. cases*	*Head circumferences in standard deviations below mean for age and sex*	*No. cases*	*I. Q.*	*No. cases*
1	14	Minus 2.0- 2.9	113	100-102	4
2	34	Minus 3.0- 3.9	51	90- 99	5
3	22	Minus 4.0- 4.9	20	80- 89	14
4	26	Minus 5.0- 5.9	9	70- 79	24
5	26	Minus 6.0- 6.9	9	60- 69	22
6	24	Minus 7.0- 7.9	3	50- 59	40
7	11	Minus 8.0- 8.9	5	40- 49	32
8	10	Minus 9.0-12.1	2	30- 39	42
9	9			20- 29	29
10	7				
11	7				
12	7				
13	4				
14	2				
15-17	9				
Total	212				

Table II. Summary of clinical diagnoses

Clinical diagnoses	*No. cases*
Chromosomal aberrations	7
Inborn errors of metabolism	8
Cerebral palsy, all forms	45
Multiple congenital abnomalies, etiology unknown*	118
Postrubella, first trimester	27
Acute cerebral infections	7
Total	212

*In this group, 8 infants with syndactyly had histories of the same anomaly in other members of the families. Many others had a variety of anomalies in the families.

corresponding to the severity of their defects. We have published similar findings.[13]

Pozsonyi and Lobb[14] reported on the relationship between mental retardation and stunting of linear and skeletal growth in 958 children. Kugel and Mohr,[15] Knobloch and Pasamanick,[16] and Casalaina and associates[17] have all published data on the association of small head size and growth failure in mentally retarded children.

We report here the results of a study of the relationship of the degree of mental retardation, as measured by I. Q., to one parameter of growth, head circumference, in children with microcephaly.

MATERIALS AND METHODS

We reviewed the records of 212 children with microcephaly who came to the Birth Defects Clinic at Stanford University Medical Center or to the Child Development Center at Children's Hospital, San Francisco, during the last 3 years. In the adult, microcephaly is defined as a condition in which the skull has a capacity below 1,350 c.c. For children we use the definition that a head with a circumference 2 or more standard deviations below the mean for age and sex is microcephalic. No children with Down's syndrome are included in the present study.

In Table I are shown the age distribution, the range of cephalic circumferences, and the range of intelligence quotients for our patients.

Ages are recorded to the nearest birthday. Only 36 were 10 years of age or older. All but 2 of the 28 cases with head circumferences of more than 5 standard deviations below the mean were found in older institutionalized children sent to Sonoma State

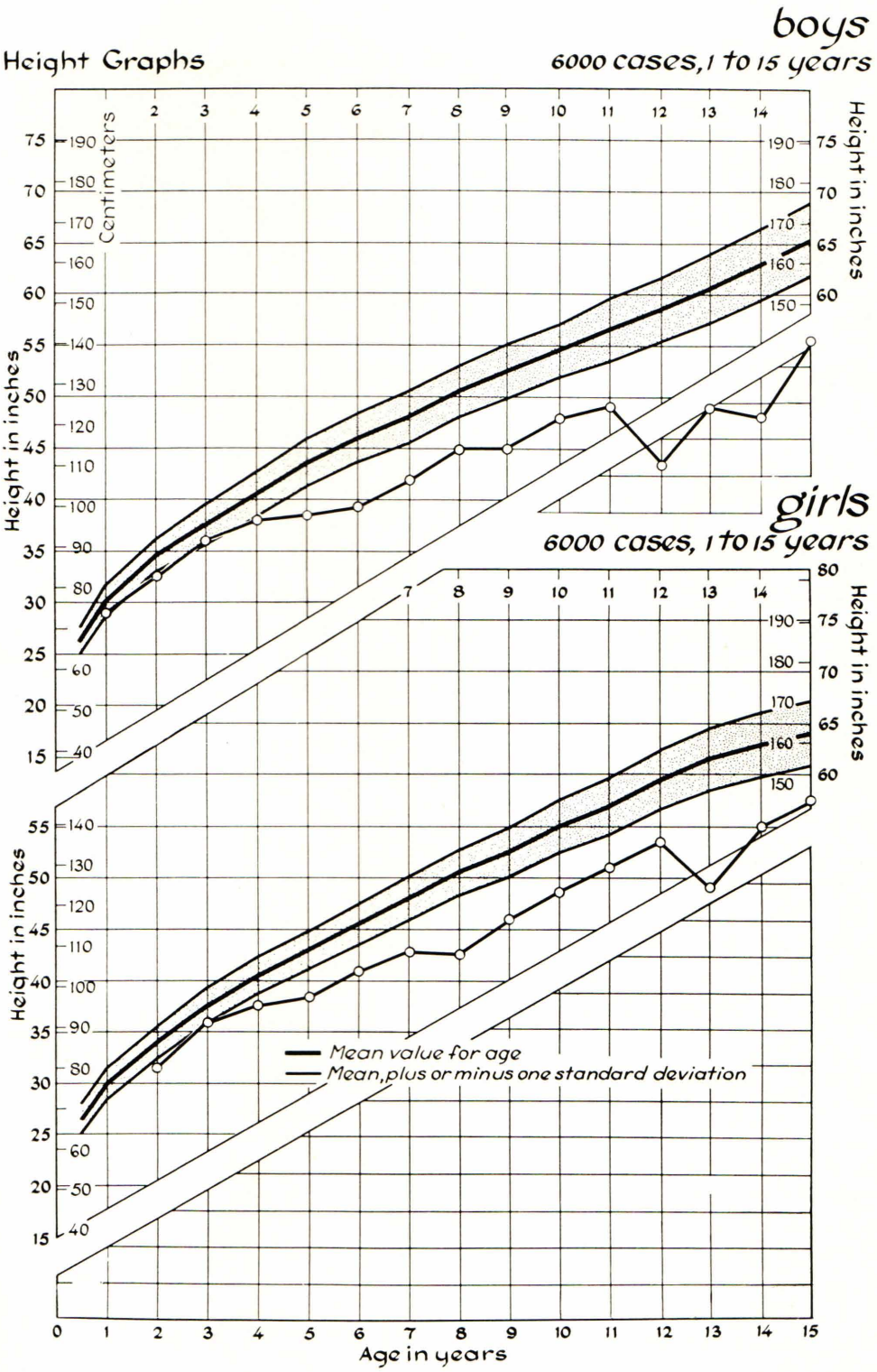

Fig. 1. Standing heights of microcephalic patients compared with normal children. Shaded areas measure plus to minus one standard deviation from the means for age and sex for normal boys and girls; O—O—O represents mean heights of 111 boys and 101 girls who were microcephalic.

Hospital, Calif. All intelligence quotients were measured by standardized, individual tests done at the two hospitals involved in the study.

Nine children, from 1 to 11 years of age, attained intelligence ratings between 90 and 102. Eight of the children in this range had cephalic circumferences from minus 2.0 to minus 2.6 standard deviations (S. D.) below the mean. The exception was a two-year-old girl with an I. Q. of 90 who had a head circumference minus 3.4 S. D. be-

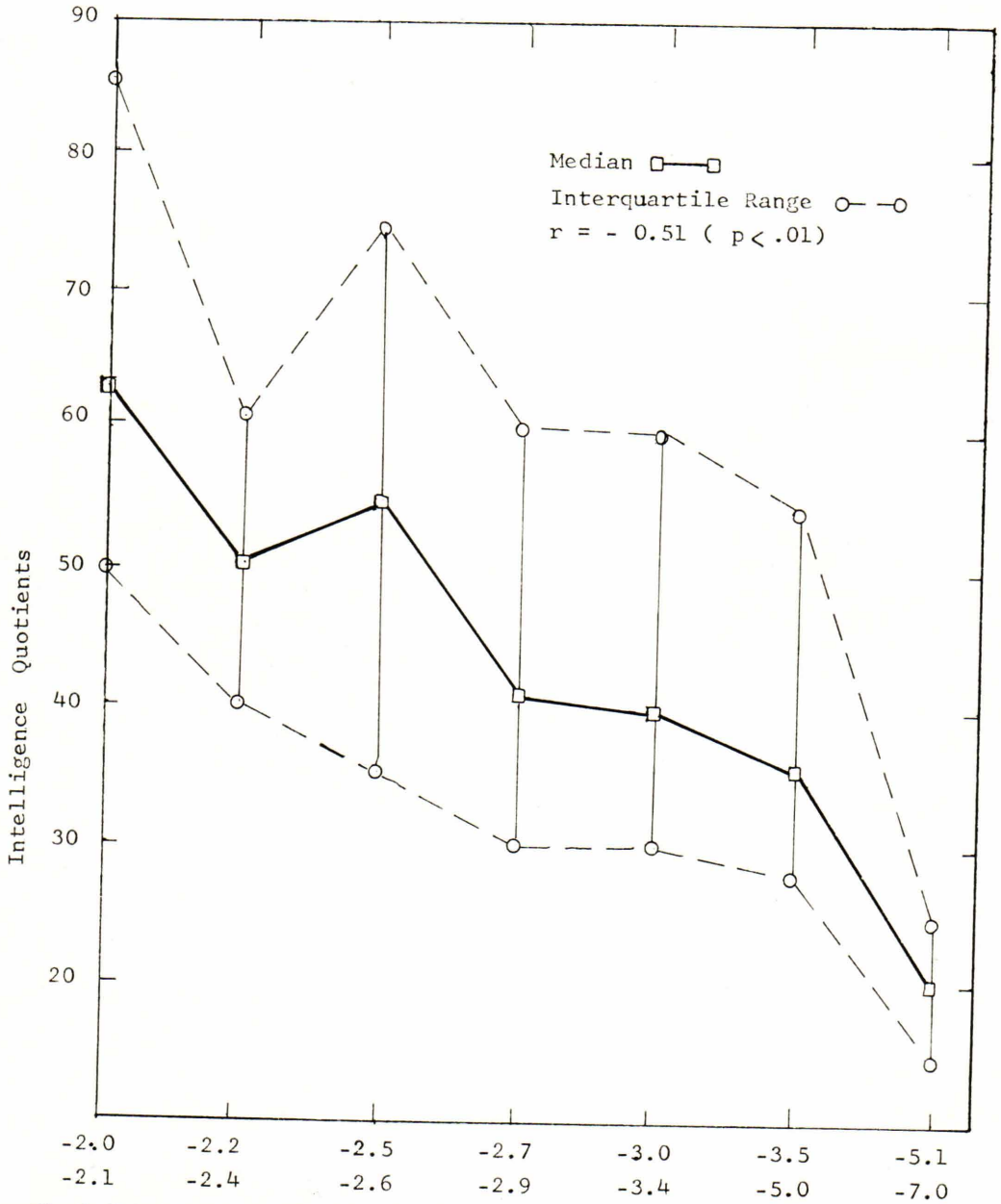

Fig. 2. Head size expressed as standard deviations below the normal means for age and sex plotted against intelligence quotients. Intervals of head circumference were chosen to include roughly the same number of observations in each group. Consequently, the intervals must be lengthened as they move away from the mean.

low the mean, and she was a congenital amputee. At the other end of the scale, all 10 microcephalic children with measurements more than minus 7.0 standard deviations had I. Q.'s of 20 or less. The 38 children with head sizes ranging from minus 4.0 to minus 7.0 S. D. had an average I. Q. of 35.6.

In the group with the top I. Q.'s, 2 children were congenital amputees, 2 had cleft palates, 2 had congenital heart defects, and 3 had cerebral palsy (hemiplegia). The latter 3 had histories of birth injuries.

Twenty-nine children had I. Q.'s below 30, and 18 of these were classified as having "primary microcephaly." Among the remainder, 2 had inborn errors of metabolism, 5 had multiple congenital anomalies, and 4 had cerebral palsy. Two of the latter had spastic quadriplegia with multiple congenital anomalies, one had ataxic paraplegia, and one had spastic diplegia with seizures. All of the group with the lowest I. Q. had more than one congenital defect. Cerebral palsy occurred among children with all grades of intelligence and all kinds of anomalies. The 3 children in the top I. Q. group, who had cerebral palsy, had no evidence of congenital anomalies or embryonic injury. Twenty-seven had histories of rubella infection before the third month of gestation. The diagnoses for the whole series are summarized in Table II. The possibility of ethnic variations was considered, but we do not have large enough numbers in the subgroups to be representative. There were 110 Caucasians, 48 American Indians, pure or mixed, 39 Negroes, 12 Orientals, and 3 of unknown parentage.

Definite stunting of growth in height is seen in our group of microcephalic children, becoming more marked as they grow older; this follows the pattern of Down's syndrome. In both groups, deficits in head size are greater than deficits in height.

In Fig. 1 the mean values for standing height of 111 microcephalic boys and 101 microcephalic girls are plotted and compared to 12,000 healthy boys and girls of the same ages in the public schools.

In Fig. 2, head size is plotted against I. Q. In the graph, medians rather than means are used for I. Q., since the data are skewed. The interquartile range is indicated by the broken lines. Since I. Q.'s below 20 could not be measured definitively, the graph ends at that point which corresponds to a head size of minus 5 standard deviations. Below this point all of our cases are institutionalized children classified as primary microcephalics. The data reveal a significant correlation coefficient of 0.51 with $p < 0.01$.

SUMMARY

Abnormally small head size has long been associated with mental deficiency. The present study relates the degree of mental retardation to one parameter of growth, cephalic circumference, in 212 cases of microcephaly. The greatest deviations from normal were found in older institutionalized children.

Intelligence quotients have a direct relationship to head size in our microcephalic patients. Some of the higher I. Q.'s were found in children whose defects could be classified as probably caused by conditions arising in pregnancy after the first trimester. The lowest I. Q.'s were found in children with chromosomal aberrations, inborn errors of metabolism, and multiple congenital anomalies. Rubella accounted for 27 cases of embryonic injury that could be ascribed to prenatal environmental insults.

REFERENCES

1. Silver, H. K., and Deamer, W. C.: Graphs of the head circumference of the normal infant, J. PEDIAT. **33:** 167, 1948.
2. Nellhaus, G.: Head circumference from birth to eighteen years, Pediatrics **41:** 106, 1968.
3. Pryor, H. B.: Charts of normal body measurements in graphic form, J. PEDIAT. 68: 615, 1966.
4. Lilly, J. C.: Man and dolphin, New York, 1962, Pyramid Publications.
5. Lilly, J. C.: Mind of the dolphin, New York, 1967, Doubleday & Company, Inc.
6. McCarthy, D.: Language development in children, *in* Manual of child psychology, New York, 1946, John Wiley & Sons, Inc.
7. O'Connell, E. J., Feldt, R. H., and Stickler, G. B.: Head circumference, mental retardation, and growth failure, Pediatrics **36:** 62, 1965.

8. Book, J. A., Schut, J. W., and Reed, S. C.: A clinical and genetic study of microcephaly, Am. J. Ment. Deficiency **57:** 642, 1952.
9. Thelander, H. E., and Goebel, G.: Microcephalia as a complication of pneumococcic meningitis, Am. J. Dis. Child. **77:** 642, 1949.
10. Thelander, H. E., and Pryor, H. B.: Abnormal patterns of growth and development in mongolism, Clin. Pediat. **5:** 493, 1966.
11. Bailit, H. L., and Whelan, M. A.: Some factors related to size and intelligence in an institutionalized mentally retarded population, J. PEDIAT. **71:** 897, 1967.
12. Mosier, H. D., Grossman, H. J., and Dingman, H. F.: Physical growth in mental defectives, Pediatrics suppl. **36:** 465, 1965.
13. Pryor, H. B., and Thelander, H. E.: Growth deviations in handicapped children, Clin. Pediat. **6:** 501, 1967.
14. Pozsonyi, J., and Lobb, H.: Growth in mentally retarded children, J. PEDIAT. **71:** 865, 1967.
15. Kugel, R. B., and Mohr, J.: Mental retardation and physical growth. Am. J. Ment. Deficiency **68:** 41, 1963.
16. Knobloch, H., and Pasamanick, B.: Mental subnormality, New England J. Med. **266:** 1045, 1962.
17. Casalaina, V., Dingman, H. F., Mosier, H. D., and Grossman, J.: The birthweight of abnormally small mental defectives, Human Biol. **36:** 229, 1964.

Letter to the Editor. When working with brain-injured children, the dichotomy of severe mental retardation versus genius among them was a constant enigma. In my extensive review of the literature[1] after retirement, I came across the following statement by Klosovoski[2] in his study of blood circulation of the brain , " The metabolic products of these cells and the waste products resulting from their disintegration (mainly their nuclei) are powerful stimulants which trigger the vigorous and massive new growth of vessels from the pial membrane into the wall of the primary cerebral vessels as observed by us during the second half of pregnancy." Is it possible that this invasion of blood vessels into an injured area may be a stimulant to adjacent tissues still in the developing stage?

I will cite two instances as examples of severely injured children followed from birth with marked physical impairment but exceptional minds. CW now twenty two years of age, a girl who graduated at the head of her class with honors and who wants to study medicine, is a severe athetoid quadriplegia with marked interference with speech. She was the first born of a young mother admitted to the hospital in labor and given a spinal anaesthia. When turned on her back for delivery, she went into severe shock, her blood pressure dropped, her pulse was barely perceptible and the physicians feared she would die. She was turned to her side, given stimulants and resuscitation. She revived and delivered a baby girl weighing 8 Lbs. 12 Oz. The infant was quiet and limp but cried when stimulated. She remained quiet in the nursery and fed poorly, however she went home with the mother, and there after was allowed in the home and office. She had no physical stigmata. At ten months she was responsive and began to react by crying to strangers, a favorable developmental sign. She was then also recognized as being athetoid. She attended the Cerebral Palsy nursery school until old enough to be admitted to a public school for the handicapped. She did not walk until 6 years of age and as noted above is still very handicapped.

The second case, NC, a boy now twelve years of age, has similar symptomatology but more severe. He is a quadriplegic, with mixed rigidity and athetosis and no speech. He also reacted with crying in the presence of strangers between nine and ten months of age. His mother maintained that he was a very bright child. He was the product of a twin pregnancy. At 5 months gestation, the mother had an episode of bleeding but carried the pregnancy past seven months. One twin was dead and from appearances, had been so for some time. The proband was described as barely viable. Dr. Morrison Gardner of the Child Development Center at Childrens Hospital, who has had many years of experience working with the handicapped, felt challanged to try to determine this childs intellectual capacity. The testing was done during many encounters and with ingenious techniques. The boy was given IQ of 140 but Dr. Gardner stated that he thought this was a conservative estimate.

The major factor in both these cases was undoubtedly hypoxia. Anoxia of more than 5 to 8 minutes is fatal. There is, therefore, a very narrow margin between survival and death. Dr. Potter[3] describes the pathology of fatal cases of anoxia as, " Sharply circumscribed petechial hemmorages most marked in the area of the brain adjacent to the ventricular system". This pathology corresponds with the symptoms of athetosis.

In discussing these findings with colleagues, I frequently get a comment suggesting that maybe he or she was brighter and 10 or 20 points lowering of intelligence would still leave a child normal or maybe above. This seems very unlikely as an explanation. I would like some comments from physicians with experience in the field,- neuropathologists, embryologists.

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References

1. Thelander, Hulda E. Interferences with Organogenesis and Fetal Development. Texas Reports on Biology and Medicine. Vol. 31. No.4. Winter 1973. pp 791-861.

2. Klosovoskii, B.N. Blood Circulation in the Brain. Translated from the Russian. Published for the National Library of Medicine, U.S. Public Health Service by the Israeli Program for Scientific Translations. 1963. pp 267.

3. Potter, E.L. Pathology of the Fetus and of the Newborn. Yearbook Publishers.

TEN YEARS EXPERIENCE WITH CEREBRAL PALSY AND RELATED DISEASES

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With ten years of experience behind us it seems timely to review changes that have taken place in the field of Cerebral Palsy in the past decade.

The staff at Children's Hospital has been exceptionally stable. Mrs. W.M. Flower, Mr. E. W. Kirk, Mrs. J. Sledge, Mrs. G. Saxton, Dr. L. Eising, Dr. E. Cabos and Dr. H.E. Thelander have been with the program either from the start or shortly after its inception. The medical consultants are essentially the same as the original group. The 1961-1962 report shows that many of the original patients are still being followed. Nearly 1000 patients have been seen which should give us a fair sampling of the problems encountered among neurologically handicapped children and their families. Furthermore, the staff members have participated actively in many and varied organizations dealing with the handicapped at local, State and National levels. Some members have visited centers abroad and many foreign visitors have been our guests. These exchanges have been very helpful in evaluating the validity of our own observations and experiences.

In general a medical problem is attacked by Research, Education and Service. The AMA Journal of June 30, 1962, has an illustration showing a patient toppling off a tripod due to the broken short service leg, the other two legs being Research and Education. The article gives the following statistics: in 1940 the Government expenditures for research were 53 million or about 7% of the total public and private medical research support. In 1961 this amounted to 496 million or 56% of the total. It is predicted that by 1970 it will reach 1.6 billion. The Institute of Neurological Diseases and Blindness of the National Institute of Health is spending millions on the causes, prevention and cure of neurological diseases in children. NIH grants include the training of personnel in the fields involved. The field of Research has obviously expanded tremendously.

The field of Education, both medical and Ancillary, has also expanded greatly. The July 1962 number of the Medical Dates Bulletin of the CMA has already listed 82 medical meetings for the months ahead. Several of the meetings pertain to subjects associated with Cerebral Palsy and its related dysfunctions. It has become almost impossible to schedule meetings without one or more conflicts. This shows an impressive interest but also poses problems. With such a large and varied number of meetings to choose from, the doctors or other participants naturally select those in which they are most interested. This means that meetings centered about a given disease entity attracts primarily those already interested. It does not reach out to the other fields of medicine where information is most needed. At a recent meeting in New York, the directors of five Development Centers from New York to California were of the opinion that to succeed in putting across the already available information on causes, prevention, and cures of neurological diseases it was essential to get the subject as well as the result of our research in some form, under some type of title, into the programs of Obstetricians, Internists, Surgeons, Anaesthetists, Radiologist, Pharmacologists, and Pediatricians. Furthermore, the subject of Child Development and the hazards peculiar to a rapidly growing nervous sytem should be taught to all medical students at the undergraduate level. This would cost nothing extra. Furthermore, since fees for refresher courses are deductable items on the income tax, refresher courses could be made self supporting. As a matter of fact, transportation and honorariums have become a means of vacation and travel for many popular speakers. This becomes an unwarrented luxury unless the patients benefit from the application of this latest knowledge.

Service to the patient has not followed the expansion pattern of Research and Education. This fact warrants serious consideration. The needs of the Cerebral Palsy patients in San Francisco are not met. This is particularly distressing since the number of Cerebral Palsy patients is not great. San Francisco is a unique county in being entirely urban and its suburbs are almost totally in other counties. The population of San Francisco has actually decreased while the neighboring counties have increased. San Francisco, with

two Medical schools until recently; with a Teachers College, and State and local institutions should have developed an ideal or model program that could have been an inspiration to growing and developing areas. With the tax funds and the private funds collected for the handicapped and available to this city, lack of money cannot be the sole cause of this situation.

Basic to adequate services are: (1.) Transportation. An excellent program is of no help to the patient who has no means of getting there. Consider a mother without a car, with limited finances, and possibly with 2 or 3 preschool children, attempting to bring a heavy, severely handicapped child across the city to a Day Care Center; or a mother who is employed trying to get even a single child transported and picked up. These are daily realities. (2) Ancillary services such as Day Care, training programs for the handicapped and recreation should be without cost to the handicapped as they are to the normal children. (3) Screening children out of existing programs without making any provision for, or taking any responsibility for children so barred from help is a deplorable situation in our communities and should not be tolerated. There should be a "home base or bases" to which such patients could be referred by the agency that turns them away and another means of help found. Existing institutions could be more elastic and accomodate the relatively few "hard to place" cases than they are at present. Unfortunately many programs start by defining what they will do and then close their eyes completely to the needs of anyone outside the limits set.

The unmet needs of the neurologically handicapped children in San Francisco and the methods by which they could be met are outlined below:

1. THE SEVERELY PHYSICALLY AND MENTALLY HANDICAPPED. (Confined to bed.) This group is not large. There are less than 100 children on the waiting list for Sonoma. Of these 100 probably not more than 20 would fall into the above category. There are probably another 20 or 25 in the same category but application to Sonoma has not been filed. From personal contact with many of these parents we know that they would utilize hospital beds intermittently if these were available and "commitment" was not demanded. The suggestion has therefore been made that the State or Local Mental Health Divisions or Sonoma (whoever is responsible) contract with the Hospitals of the city to hospitalize these patients in the regular Departments of Pediatrics until a transfer to a State Institution can be made. Since these patients have already been carefully diagnosed, hospitalization could be on a minimal care basis. Pediatric Departments tend to have an occupancy of 50% or only slightly over, beds and staff therefore are available. Although the number of these patients is not great, the hardship to the family is disproportionately grave.

2. DAY CARE AND OCCASIONAL BOARDING. (Patients screened out of all educational and training institutions.) If Day Care services were free and transportation provided there might eventually be 75 or possibly 100 children so cared for in the City. The exact number could not be determined without a trial. There are some parents who prefer to take care of the children at home. There are other children whose training status can only be determined by trial. There are a few children in existing facilities that might be more suitably taken care of in a Day Care situation. If provision were made now for 20 or 25 children it might take some time to fill the quota. It would be prudent therefore to start with a relatively small activity and provide for expansion if and when this becomes necessary. This is again a problem that concerns a relatively smaller group but one that poses disproportionately grave situations.

3. THE MULTIPLY HANDICAPPED. The past year Sunshine School provided two additional classes; one for the blind-neurologically handicapped and one for the hyperkinetic neurologically handicapped. There were ten children thus accomodated. We hope

these classes will be continued. One of our children who is deaf and hyperkinetic was sent to Napa State Hospital for lack of other residential facilities. A great deal of work by several agencies was done on this family, all of whom recognized that Napa State was not the appropriate facility but it was the only one available.

4. UNDER 8 YEARS, POINT TWO AMBULATORY CHILDREN. A class for 10 or 12 children between the ages of 5 and 8 years at Louise Lombard School would take care of a group of children not accepted in Sunshine School because the degree of their motor handicap in their opinion does not justify admittance. The Point II child is easily diagnosed before 8 years of age. It is well recognized that early training for these children is extremely important. The parents also need guidance in home training before 8 years of age.

5. AID TO RETARDED CHILDREN NURSERY SCHOOL. This facility accomodates some of our children who have become ambulatory and who are not eligible for Sunshine. More parents could use it if transportation were provided and if mother participation were not required. Working mothers and some others cannot always arrange for participation.

6. APHASIA AND DYSLEXIA. Children with major or minor physical handicap but with aphasia or dyslexia pose a serious problem in this area of the country. The State Cerebral Palsy School with generous tax support remains a diagnostic and evaluation center and is of very limited value as an educational institution. A child with aphasia needs years, not weeks or months, of education. With the expansion of diagnostic centers throughout the State the function of this institution should be re-evaluated. The function of the Deaf and Blind State Schools should also be re-evaluated. At present they will not take neurologically handicapped deaf or blind children. Inasmuch as provisions for the education of the "normal" blind and deaf have expanded the question might well be asked "Do we need residential schools for "normal" blind and deaf children?" If some of these children come from remote or isolated areas, could they not board in a normal home and attend school in a nearby urban place? It seems to us that these institutions might need to change with the times and provide more urgent services such as long term training for the aphasic, the severe dyslexic, the blind and deaf neurologically handicapped and the adolescent severe cerebral palsy. Stanford Medical School is launching a project for aphasic children; being in a medical school it will probably be able to give only limited service.

7. PREPARATION FOR USEFUL LIVING. Our experience indicates that our schools are too academically oriented. The pupil finds himself at 16 or 18 years of age out of school and without any concept what he is to do next. This is late to turn to rehabilitation. Actually for the child with a permanent physical handicap orientation toward a realistic work goal should start at 12 years of age and the schools should be so geared. From the experience in sheltered work shops I.Q's by standard testing are not the total indication of ability to perform in a work program. Since most of the children are in need of practical rather than academic training, special education should be directed toward work performance when the children reach the age of 12 or 13 years.

The professional concept of Cerebral Palsy has changed in the past decade. This is aptly illustrated by the British who in January of this year changed the name of the "Cerebral Palsy Bulletin" to "Human Development and Childhood Neurology". President Kennedy in defining Mental Retardation to his committee for the study of the subject made that definition so broad that for practical purposes it covers 75% or more of the Cerebral Palsy cases. At the local level, Dr. Kimmicks committee on Mental Retardation accepted President Kennedy's definition as a working definition. The preliminary tenor of the above two groups (President's committee - Short Doyle Funds) is toward service to the patient. We can only hope they are not deflected from this ~~goal~~ course put in Capital,

Also in the past decade there has been at least two new organizations in the field, one for the Neurologically Handicapped and one for the Multiply Handicapped. Groups such as these are formed because services are not available to the children. If the existing organizations supplied the services new parent groups would not have been formed. On the other hand there has been a tendency also for parents of handicapped children to unite. "Concern" is in part such a group. At the local level there has been at least two joint meetings of parent groups, and a third is contemplated. Parents are not the only ones seriously concerned about the lack of services, donors increasingly are disturbed by the multiple demands for their charity dollars. To both parents and donors the super structure seems to grow and expand becoming more complicated and more remote from and with less services to the patient.

The cartoon cited earlier pictured the patient falling off the tripod due to the short broken service leg, it is axiomatic, however, if services break down the super structure will topple eventually too.

A great deal has been learned as a staff working intimately with patients and families, some now for ten years, as well as by exchanging experiences with others doing the same type of work. There is general agreement as to what is needed. Surveys, demonstrations, and pilot studies are passe in the field of the neurologically handicapped. Temporary grants are a delusion; steady continued endeavor is needed. Programming for these children is neither extremely difficult nor unduly expensive. The focus must be on the patient and his immediate family; and then the skills of the different staff members must be effectively utilized. This means recognizing and respecting the roles of every member of the program. It is an Orchestral rather than a team approach.

Recognition of human worth needs to be re-emphasized. It has happened time and again when parents have a satisfying diagnosis and interpretation of their problem and some support in dealing with it, they in turn interpret it to their neighbors, the church, and the school. Many families have found ready help and cooperation all around them. When people begin to help people there is not much need for a complicated and expensive super-structure.

Although progress has been slow and often discouraging the past ten years, the momentum appears to be increasing and the next ten years should show far greater accomplishments.

H. E. Thelander, M.D.
With the assistance of
Mrs. Gertrude Saxton, Social Work Consultant
Mrs. Jane Sledge, Nursery School Director

CHILDREN'S HOSPITAL OF SAN FRANCISCO

Child Development Center — Statistical Report — January 1 - December 31, 1975

BREAKDOWN OF SOCIAL SERVICE STATISTICS*

Age	% Male	% Female
0-5	20.22	9.55
6-11	47.75	14.61
12-16	3.37	4.49

Family Status	%
Single Parent	40.45
Two Parents	51.12
Foster Parents	2.25
Guardian	6.18

Race	%
White	53.93
Black	26.40
Spanish Surname	10.67
Japanese	Ø.ØØ
Chinese	2.25
Filipino	4.49
American Indian	0.56
Other Non-White	1.69

Area	%
Eureka - Noe	9.55
Westside	20.22
Southern	19.66
Northeast	5.06
Sunset - Richmond	21.91
Non-San Franciscans	23.60

Referral Source	%
School	48.88
Physician	28.65
Dept. Social Services	1.69
Dept. Public Health	8.43
Children's Hospital Clinics	3.93
Self	5.06
Other	3.37

Pay Category	%
100%	28.65
80%	3.93
60%	5.62
40%	13.48
MEDI/CAL	42.13
CCS	3.37
MEDI/CAL + CCS	0.56
AHCP	2.25
Full Insurance	Ø.ØØ

Educational Level	%
Pre-School	19.10
Kindergarten	10.67
First Grade	17.42
Second Grade	11.24
Third Grade	12.36
Fourth Grade	6.18
Fifth Grade	6.18
Sixth Grade	2.81
Seventh Grade	0.56
Eighth Grade	1.69
Ninth Grade	0.56
Tenth Grade	1.12
Ungraded	3.93
EH Class	1.69
EMR Class	1.69
LDG Class	2.81

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TOTAL NUMBER OF VISITS PER SERVICE

Neurology	498
Psychology	395
Psychiatry	37
Social Service	340
Pediatric	163
Occupational Therapy	1170
Physical Therapy	1170
Educational	25
Nursery School	1454
Orthopaedic	33
Audiology	4
Amputee Clinic	24
Total Visits	5313

*Of a total of 232 new patients seen in Social Service, statistics are available for only 178.

LF:mls
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