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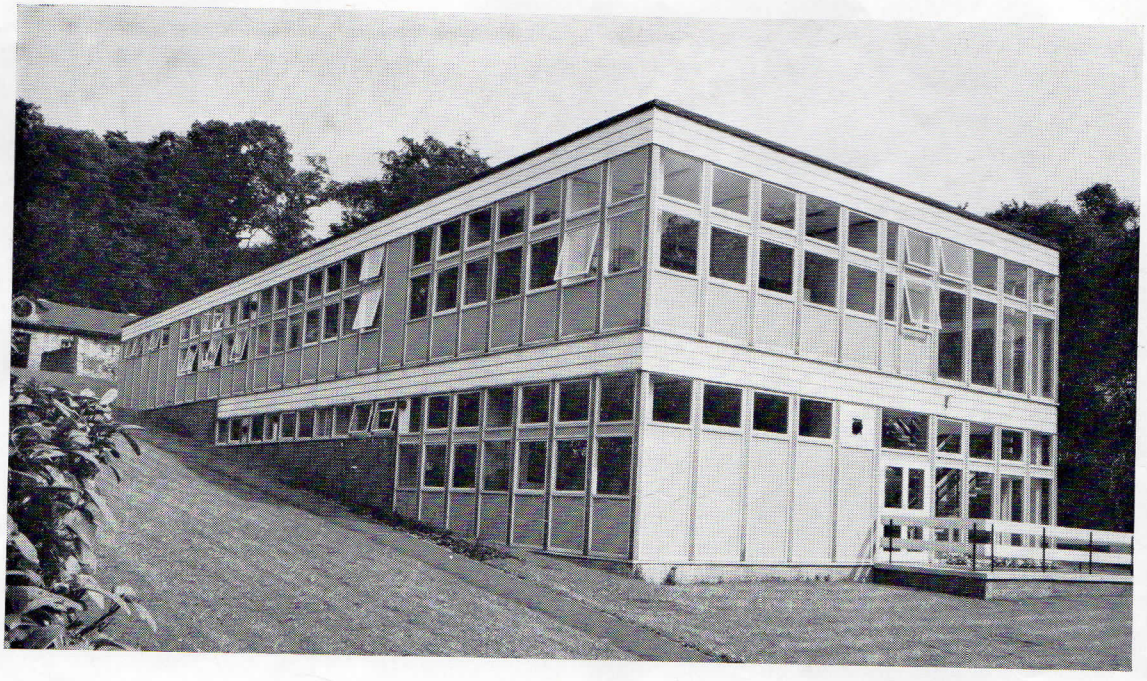

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During the past decade, the investigation of spina bifida and hydrocephalus has constituted a major part of the research programme in the Department of Child Health and the Children's Hospital in Sheffield. It was the need for further intensive research into these conditions that prompted the Department to seek the means to establish additional accommodation and equipment. This article, which describes and illustrates the work, equipment and aspirations of the new unit, acknowledges the invaluable financial assistance of Sir Isaac Wolfson, The University Grants Committee, The Hospital Endowment Fund, The Richard Fund, Action for the Crippled Child and many groups and individual donors.

THIS unit* is primarily designed for the study of spina bifida and hydrocephalus which are among the commonest congenital malformations. Thus each year some 2,500 infants in Great Britain are born with one or other (or both) of these disorders and some three-quarters of these are liveborn. In addition, about half this number are born with anencephaly, and although all these latter infants are stillborn or die shortly after birth (and thus present no long-term clinical problem), aetiologically their malformation is closely related to spina bifida. Indeed, the two conditions frequently occur in the same sibship and therefore anencephaly is of great research interest.

*Opened 28 February 1968

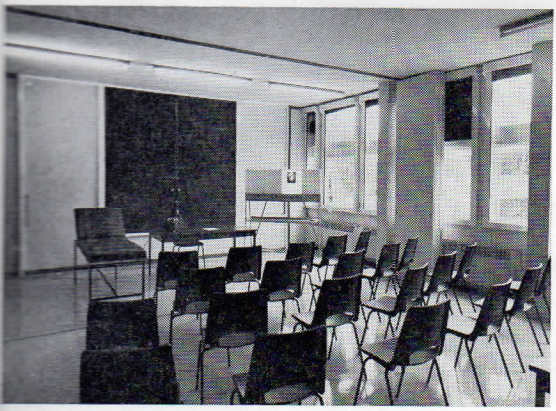


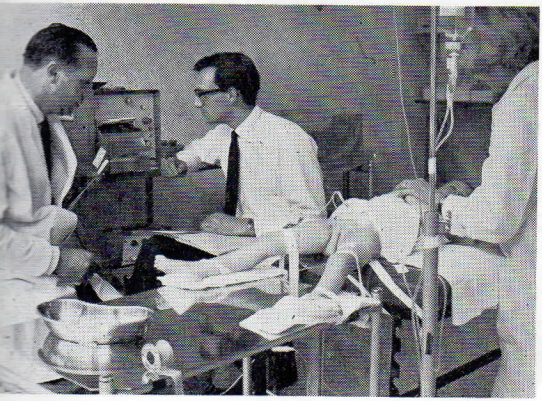
Fig. 1 *Lecture theatre in the Unit.*

Fig. 3 *The histology laboratory.*

Spina bifida is a condition in which part of the spinal cord has not been properly formed and its normal coverings of soft tissues, vertebrae and skin are also deficient, so that part of the spinal cord lies exposed on the surface of the baby's back at birth, being covered only by a thin layer of meninges. Most commonly, the lesion is in the lumbar or lumbosacral region but occasionally it is much more extensive. In other instances, the lesion is higher up. If it occurs at the base of the skull, it is called encephalocele. The sac of the encephalocele may be as large as the skull itself and it often contains abnormal brain tissue. This type of lesion is most closely allied to the most severe lesion, anencephaly. The sex incidence of encephalocele and of anencephaly is about the same—seven girls to three boys. In spina bifida, parts of the spinal cord may be maldeveloped and abnormal before birth, leading to paralysis or weakening of the legs and of the muscles of the bladder and the rectum. Much additional damage can be caused to the cord by exposure, drying out and infection but this is largely preventable by operation on the first day of life. This fact was established by our research team and, subsequently, operation on the first day of life has become standard practice in most centres in the western world.

Hydrocephalus of obstructive type is present in at least 80 per cent of cases of spina bifida. As has also been shown in our Unit, this hydrocephalus is present at birth and does not result from operative closure of the spina bifida. It can vary from trivial to extreme severity. Untreated and untreatable hydrocephalus in children with

Fig. 2 *Bladder pressure studies in progress.*



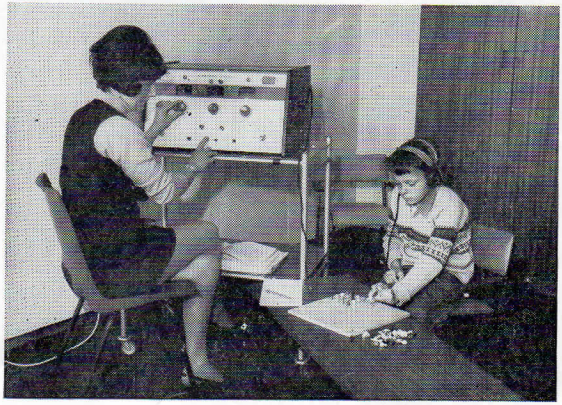
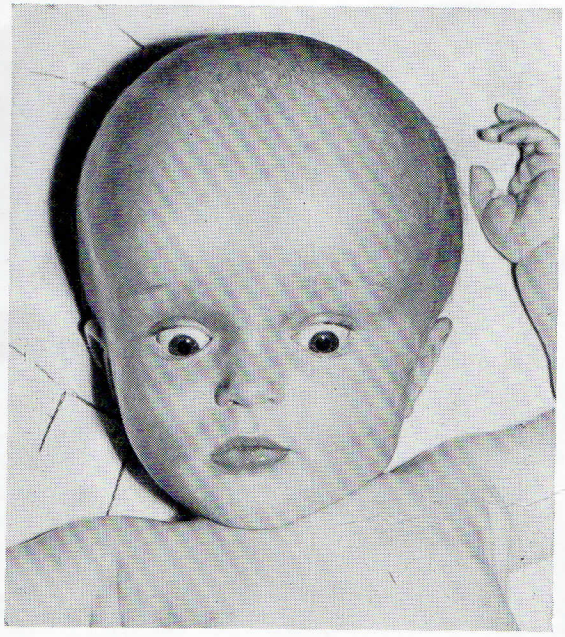


Fig. 4 *Audiometry in progress.*



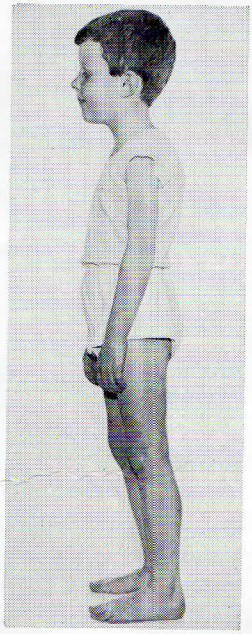


Fig. 5 (*left*) *Frontal view of baby with well-marked hydrocephalus before operation.*

Fig. 6 (*right*) *The same child at age 7 years.*

spina bifida used to carry a high mortality, and few of these babies survived to school age. Hydrocephalus unassociated with spina bifida also carried a grave prognosis, and although more of them survived, most survivors were spastic, blind, mentally defective and subject to convulsions. The head used to attain a tremendous size.

Another major advance, introduced about 12 years ago, was the more effective treatment of hydrocephalus, by effecting a reduction of the increased intracranial tension by bypassing the obstruction in the cerebrospinal fluid pathways in the brain. In principle, the technique consists of implanting a plastic tube to run from the cerebral ventricles to the right cardiac atrium. In the Holter procedure, one end of the tube is carried subcutaneously behind the ear and is attached to two uni-directional slit valves, each housed in a metal casing, and separated by a short length of compressible plastic tube. The other end of the tube runs from the valve assembly into the right cardiac atrium through the jugular vein and the superior vena cava. A "reservoir", developed by P. P. Rickham, can be inserted into the system at the point at which the ventricular catheter reaches the surface of the brain or the valves may be located elsewhere. None of these shunts or shunting procedures is perfect and the complications, which tend to occur even many years after insertion, are numerous. Much research is needed, and possibly quite different methods of treatment from those in current use today may emerge.

Meningitis is another major hazard for infants born with spina bifida. This usually occurs in the first weeks of life and is commonly related to infection of the spina bifida site and to the problem of wound closure. Meningitis may occur in as many as 20 per cent of such infants treated and, even without spina bifida, neonatal meningitis still carries a very high mortality and serious sequelae may occur in the survivors. The presence of overt sepsis and an exposed spinal cord greatly adds to the hazards. In addition, meningitis in these cases is often caused by unusual organisms such as *Bacillus proteus* or *Pseudomonas pyocyanea*, which used to defy existing antibiotic drugs. Major advances in treatment have been achieved in recent years by fully exploiting the potentials of newer antibiotic drugs and by the systematic study of drug concentration in the cerebro-spinal fluid in hydrocephalic infants. These concentrations must exceed the minimum inhibitory concentrations of the drug against the organism causing the disease. In a large series of patients studied in this Unit, it was found to be impossible

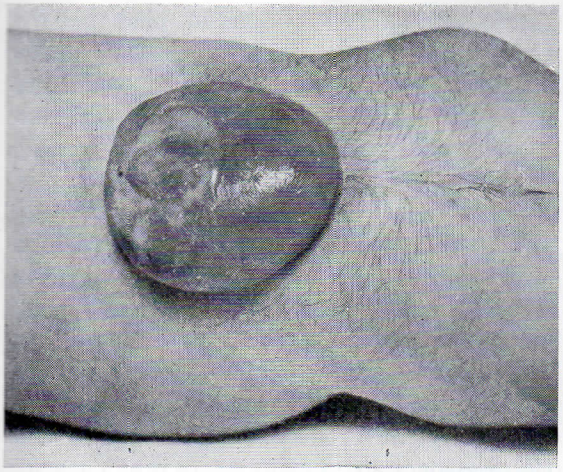


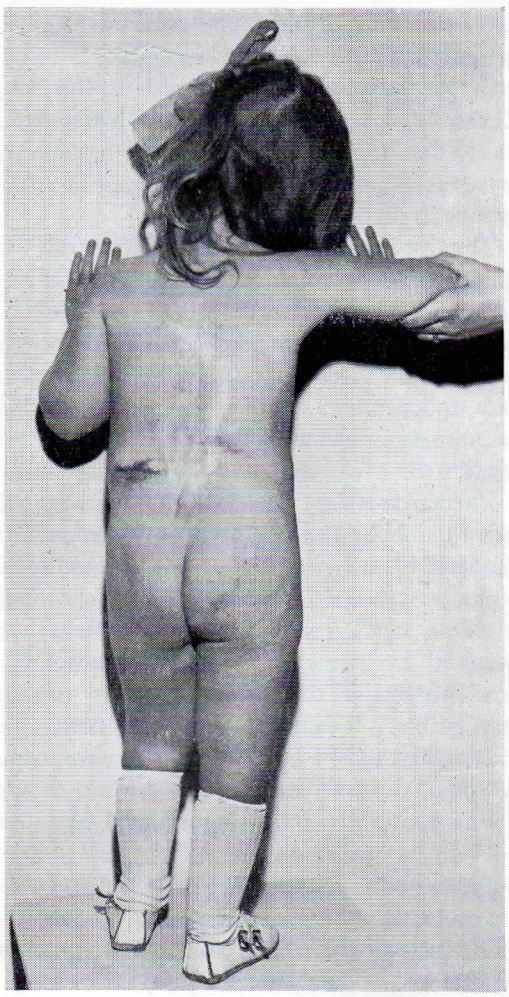
Fig. 7 *A typical lumbosacral spina bifida before operation.*

to reach adequate levels without intraventricular injections. The study of these problems, as well as the study of the prevention of infections, is a major part of the work covered in the new Unit.

Another important disability in children whose spina bifida is in the lumbar or sacral area is dribbling incontinence of the bladder with the triple risk of recurrent and chronic pyelonephritis, of progressive dilatation of the lower and upper renal tract and of major social handicaps. The first two risks combined may lead to progressive renal failure, uraemia, hypertension and death from renal causes, which is a major hazard during the later years of childhood. In this field much research is necessary to determine the value of any promising drug in the prevention of infections and the treatment of established infections. There is a need to determine the value and timing of operative procedures, such as dilatations of the bladder neck and of bypass operations. The prognostic value of bladder pressure studies in early infancy, and of pressure studies in ileal loop diversion, has yet to be established. The Unit is fully equipped to carry out such investigations, and these are in progress. Today, there are no longer infections which cannot be treated by appropriate drugs, though the problem of preventing new infections, relapses and reinfections is far from solved.

Orthopaedic problems do not threaten life but are nevertheless of great importance. Paralysis of various degrees and deformities are very common and can make an intelligent child's life a severely restricted one. Intensive research in prevention (early operation) and the treatment of paralysis has resulted in a substantially improved outlook for these children, nearly all of whom will be able to walk. The discovery and development of the ileo-psoas transplant for the treatment of neurogenic dislocation of the hip

Fig. 8 *A child five years after an operation for an extensive thoraco-lumbar meningomyelocoele. Although there is considerable scar formation, the child is able to stand.*



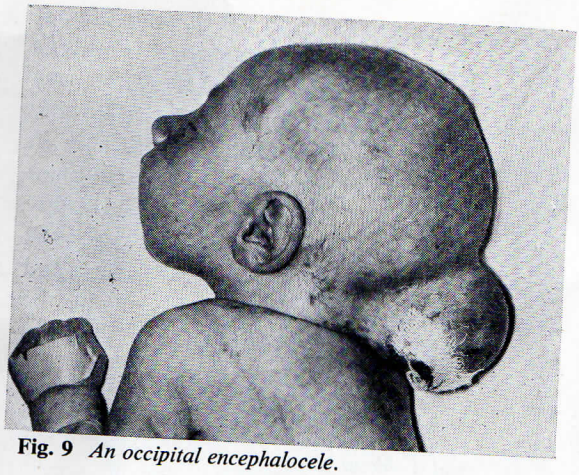


Fig. 9 *An occipital encephalocele.*

and osteotomy of the spinal column to prevent extreme kyphosis are only two among the major orthopaedic advances developed in this Unit. Currently, detailed studies are now being carried out to determine the prognostic value of electromyography and of faradic responses in muscle groups in the lower limbs.

The five main fields for clinical research—the spinal lesion, problems connected with meningitis, hydrocephalus, problems of the renal tract and the orthopaedic aspects—could not be properly pursued without major contributions and independent studies in pathology and bacteriology, including the experimental production of spina bifida and of hydrocephalus.

Further investigations into the aetiology, the risk to siblings and the potential future risk to children of surviving spina bifida and hydrocephalus subjects are essential, as knowledge in these matters is far from complete. For example, in spite of a considerable amount of established data, there is no unanimity even on such points as whether spina bifida is determined by heredity, or whether it is due to environmental causes or to some combination of these factors.

Finally, results and progress must be judged not only by survival figures and physical progress but also by the intellectual and social success of the patients and the assessment of the total integration of the person, and his usefulness to himself and to the community.

It is with all these aspects in mind that th Unit was designed. The upper floor hold administrative offices, a lecture and conferenc room with a library, a data processing unit an consulting rooms for special study. There is soundproofed room for audiometry, a psychologist's office (with one-way viewing window and an orthopaedic assessment unit. There is also a theatre suite, in which minor operative investigations can be carried out, such as bladder pressure studies using electronic techniques; it also accommodates an echograph, an apparatus for studying uptake of radioactive substances and equipment for electroencephalography and electromyography. Finally, there is a "clinical" laboratory for simple, everyday investigations.

The lower floor is largely occupied by the Unit of Research and Experimental Pathology and is well provided with equipment through the generosity of the University Grants Committee. On this floor also are the two rooms of the Photographic Department.

Apart from the University Grants Committee, which provided the bulk of the equipment, scientific and office equipment has been and is being provided by the Richard Fund, which was specially constituted for the promotion of research into spina bifida and hydrocephalus at this Unit and at the Children's Hospital in Sheffield. The Fund also provides salaries for research, technical and office staff. Other major support is given to the Unit by Action for the Crippled Child, who support two major projects by providing equipment and salaries for research and secretarial staff. Much help has also been received from manufacturing firms who support specific studies with staff and free supplies of drugs.

Funds permitting, the unit could accommodate many more research workers. The number of possible projects is almost inexhaustible, and the case material available for study of all kinds is virtually unlimited. Some 200 cases a year of spina bifida and/or hydrocephalus are dealt with and there is clinical or pathological material relating to well over 1,500 cases.

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