MEDICAL PRACTICE

Contemporary Themes

Life with spina bifida*

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Spina bifida is a broad term that includes minimal lesions such as spina bifida occulta and cystic swellings not containing any neural tissue (spina bifida with meningocele). Our main concern, however, is spina bifida with open myelomeningocele, in which the neural tissue is exposed on the surface of the cystic swelling.

The neural tissue in this exposed plaque is abnormal and defective, so that the muscles innervated by it, and indeed distal to the plaque, may be partly or completely paralysed. Naturally, innervation of the bladder and bowels is almost always defective.

In addition, about 90% of the children with open myelomeningocele have hydrocephalus, but the degree of ventricular dilatation and intracranial tension varies and is not directly related to the appearance of the back lesion. If an infant had an operation to close the back wound in the neonatal period he might require a valve for the hydrocephalus within the first six to eight weeks. Complete paralysis of the legs may lead to no deformity whatever, but partial paralysis may lead to dislocation of the hip and to foot deformities. If it is decided to encourage the child to walk with appliances several operations may be needed on the feet and also a muscle transplant at the hip to keep the hip in joint after reduction. The other serious orthopaedic problem is the spinal deformity caused by unopposed action of certain muscles, which leads to kyphosis or kyphoscoliosis and which may require major surgery for partial correction.

In treating the hydrocephalus the excess ventricular fluid can be shunted through the Holter valve back into the bloodstream,

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and although some patients require revision of the valve several times in the first ten years, the hydrocephalus can be reasonably well controlled. We are learning in Sheffield how to prevent, and also recognise and treat, infection of the valve system; but as with any foreign body, the risk of infection remains.

Defective bladder emptying may lead to stasis of urine within the renal tract and a tendency to infection, and in certain cases may lead to high pressures that cause the upper tract to dilate. Careful and frequent monitoring of the renal tract has enabled us to detect the early signs of trouble and give appropriate treatment, and there is no doubt that young children under treatment now have better kidneys than did children ten years ago. Even so, a ten-year review of survivors of an unselected series showed that nearly a quarter of them had normal or nearly normal bladder control.

Expression of the bladder every two hours may enable the incontinent to be socially dry; but, if not, a penile appliance will help the boys, and an indwelling Foley catheter has been a tremendous boon in many girls. We still, however, have to undertake a urinary diversion in many of these patients.

A typical 10-year-old with severe spina bifida will therefore spend most of his time in a wheelchair but can make some progress in long calipers; his hydrocephalus is controlled with a valve that may have needed two or three revisions during the growth period. Most children have an IQ within the normal range but below the mean; intellectual attainment will depend on the degree of hydrocephalus at the time when treatment was started, whether there has been infection, the speed with which obstruction has been relieved, and the opportunities for intellectual stimulation. I noticed a remarkable change in the children when the spina bifida school was first opened in Sheffield, because many were receiving stimulation that had not been possible before. A boy will probably have a good upper renal tract and may be wearing a penile appliance if he cannot be kept dry by expression of the bladder. A girl may have an indwelling Foley catheter or may have needed an ileal conduit. This is not an unusual picture of the young person with spina bifida and hydrocephalus. I would emphasise that this is a

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person who has spina bifida, and it is very important that we always refer to them and treat them as persons. The picture at 15 or 20 years is unlikely to change significantly, unless there is an untreated obstruction of the valve system or an uncorrected stasis and back pressure in the renal tract.

Should they be allowed to die?

At the end of the 1960s my medical colleague Dr John Lorber became very concerned about the degree of handicap of these young people and the morbidity and hospital admissions that resulted.¹ His extensive study showed that those who have a gross lesion at birth are likely to be severely handicapped when they grow up. He came to the view that the degree of handicap could be forecast at birth, and became strongly opposed to any treatment at all of severely affected babies in the newborn period. He proposed certain criteria for selecting those infants who should be allowed to die, and these were widely accepted—partly because of the human appeal of the view that these children should not be allowed to suffer, and that their parents should not have the burden of looking after them. It was also attractive to administrative authorities because the burden of the financial support needed to treat and care for children with spina bifida would be greatly reduced.

The cardinal error in these proposals is the expression, they should be allowed to die. Thirty years ago I was concerned with the care of many patients with spina bifida at the Children's Hospital in Boston, Mass, under Dr Franc Ingraham. The policy was that *none* of these children should have a neonatal operation. The lesion was covered by a simple dressing and protected by a ring; and after about a week or ten days the children went home to be looked after by their parents and the local paediatrician, with monthly visits to the neurosurgical and orthopaedic clinics. I have seen many children with severe lesions that epithelialised spontaneously, who then had the operation to close the back wound at about 18 months. Several of the very severely affected ones died at home, but I myself have handled many survivors with serious lesions. I thought it was an unduly heavy burden for the mother to have to look after the swelling on the back, and when I first came to Sheffield I decided to remove the swelling in the neonatal period if possible. Even so, for several years I continued to see children with quite obvious severe lesions who had not undergone any operation immediately after birth.

There is a widespread myth that if you operate on a child with spina bifida the child will live, and if you do not operate he will die. This is nonsense. They will not all die spontaneously.

"No treatment" methods

How is it then that those who write about the value of selection can point to a high mortality, usually 100%, in those that they have selected out? I am sure it has nothing to do with the administration of antibiotics, because those young patients in Boston had no antibiotics 30 years ago. We must look at the exact method of management of those who have no treatment.

One paediatrician has said that they receive the same care and attention as any other baby, being fed and picked up and loved, and even taken out into the sunshine in a pram. Yet these babies are receiving 60 mg/kg body weight of chloral hydrate, not once but four times a day. This is eight times the sedative dose of chloral hydrate recommended in the most recent volume of *Nelson's Paediatrics* and four times the hypnotic dose, and it is being administered four times every day. No wonder these babies are sleepy and demand no feed, and with this regimen most of them will die within a few weeks, many within the first week.

It is sometimes said that the chloral hydrate is being administered for pain, either in the back or due to the hydrocephalus, but I personally have seen little evidence that the babies have

pain in the newborn period, nor have I found them unable to sleep. And if chloral hydrate is such a good drug why is it not given to those who are intended to survive?

There is nothing secret or confidential about this method of management, for the paediatrician has stated that he has published the details of management for all to see. At a meeting of doctors, social workers, and theologians another paediatrician explained his method of asking the registrar to administer morphine. He claimed that the parents were taken fully into his confidence and were 100% behind him. When closely questioned he admitted that he did not tell them that the child would receive morphine—merely that the child would die.

In another centre only one out of 24 patients was operated on—all the others died. When asked, "Did they fall or were they pushed"—into death—the reply was, "They were pushed of course." At another meeting I attended a paediatrician was asked by a medical student what was his method of management, and the reply was, "We don't feed them."

I have mentioned the various problems that a person with a moderately severe spina bifida might have in his life, and emphasised that the duration of life would depend on the attitude of the doctor in charge. Some have been prepared to say in public that they would administer drugs, whose direct purpose and effect is the death of the child in a relatively short time. I think there are some paediatricians who have misunderstood the likely consequences of simply withholding surgery, and I have had to deal with children at 6 months of age in whom the total drug treatment part of the regimen had been omitted. I have indeed treated some who have had chloral hydrate, later phenobarbitone, and later morphine and have still survived to require operation. Such doctors are entitled to their view that their actions are best for the child and family, but there should not be any pretence that all these babies are dying spontaneously. Indeed, one must ask, "Are not these actions outside the law?"

I think it is extremely important from a strictly scientific point of view that we cannot regard the non-operative mortality figures as indicating accurately how many babies who are selected for no treatment will die spontaneously—there is, so to speak, an inbuilt insurance policy. There is one further worrying aspect. The editors of medical journals are conscious of their responsibility not to publish papers that are clearly unethical—for example, reporting research that required unnecessary operation or any appreciable risk. How much more carefully should they look at contributions based on studies that include the administration of drugs to accomplish the death of a child?

Criteria for operation

It may be asked whether I would advocate operation on every baby with spina bifida. Of course not. As with every aspect of surgery, there are criteria for selection, which should be based on sound medical and surgical principles and a knowledge of the prospects with and without surgery. My scheme for selection for operation in open myelomeningocele is as follows, babies in categories 1 and 2 having no operation.

Category 1

This category includes those patients who are judged likely to die within a few days or a week or so—for example, babies with severe intracranial haemorrhage or another major life-threatening anomaly. There will be no operation on the back in these children because it could have no bearing at all on whether they lived or died.

Category 2

Some of the children who seem unlikely to die spontaneously in a short time will have a serious lesion of the back—for example, a very wide lesion or one producing a severe kyphosis, in which the chances of primary healing after surgery would be small: there would be a risk of wound breakdown, which would be far worse than no operation at all. Epithelium is likely to grow over the exposed plaque with simple protection of the wound, and this would be the method of treatment.

Category 3

Other babies are judged to have a good chance of primary wound healing after operation, and these are placed in three grades of severity.

Grade 1—Active movement of the legs has been observed after birth. These babies need urgent operation because otherwise some of that active movement may be lost.

Grade 2—Babies in the intermediate group have certain active muscles such as hip flexors and adductors, and perhaps quadriceps. I think these are important muscles to preserve for the orthopaedic surgeon to use, and I would advocate urgent surgery.

Grade 3—At the other extreme there are babies who make no observed leg movements after birth; in these infants operation on the back will have no effect upon the muscular power in the legs. They can be treated either by a non-urgent operation (but within 24 to 48 hours to minimise infection) or by conservative means such as simple dressings.

The survivors, whether they have been operated on or not, will be offered surgery for hydrocephalus or for renal tract or orthopaedic problems to improve the quality of life or prevent deterioration.

Under no circumstances would I administer drugs to cause the death of the child. There is no doubt that those who are severely affected at birth will continue to be severely handicapped. But I conceive it to be my duty to overcome that handicap as much as possible and to achieve the maximum development of their potential in as many aspects of life as possible—physical, emotional, recreational, and vocational—and I find them very nice people. If in the *neonatal* period you are looking for opportunities to make sure they will die I am sure this attitude is likely to persist even when they are older. Some have been regarded as living completely miserable and unhappy lives.

Yet when I see them I find them happy people who can respond to concern for their personal welfare.

One final point. It has often been said by those who oppose abortion that the disregard for the life of a child within the uterus would spill over into postnatal life. This suggestion has been "pooh-poohed," yet in spina bifida there is a clear example of this. The equanimity with which the life of a 17-week-gestation spina bifida infant is terminated after the finding of a high level of α -fetoprotein in the amniotic fluid has, I think, spilled over to a similar disregard for the life of the child with spina bifida after birth.

Much of my surgical life over the past 30 years has been devoted to improving the quality of life of those who have been born with spina bifida. The attitude of mind that would eliminate all the severely handicapped reminds me of the poster issued by Christian Aid some years ago, which said "Ignore the hungry and they will go away"—to their graves. If we eliminate all the severely affected children with spina bifida there will be no more problem; but why stop at spina bifida, why not all the severely affected spastics, all those with muscular dystrophy, and all those with Down's syndrome? Why stop at the neonatal period? Our aim should be that life with spina bifida is the best possible life for that person in the family and in the community.

Reference

¹ Lorber, J, *Developmental Medicine and Child Neurology*, 1971, **13**, 279.

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Clinical Topics

Management of airway complications of burns in children

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Summary

Children who have been exposed to smoke in a confined space or who have soot or burns, however minimal, on the face should be admitted to hospital. Respiratory distress may be delayed, but if it is progressive the patient should be curarised, intubated, and mechanically ventilated. Unless ventilation continues for 48 hours, followed by 24 hours' spontaneous respiration against a positive airway pressure, stridor and pulmonary oedema may recur. An endotracheal tube small enough to allow a leak between it and the oedematous mucosa must be passed to prevent laryngeal damage and subsequent subglottic stenosis. High humidity of inspired gases keeps

secretions fluid and the endotracheal tube patent. A high oxygen concentration compensates for deficient oxygen uptake and transport caused by pulmonary lesions and the presence of poisonous compounds interfering with oxygen transport. Dexamethasone to minimise cerebral oedema and antibiotics to reduce the incidence of chest infections should be given.

Introduction

It is increasingly recognised that accidents producing burns or scalds may also damage the respiratory tract, and that this damage may be much more serious than the extent of the surface injury would suggest.

A pulmonary syndrome caused by inhaling hot irritant fumes was first described in 1942¹⁻⁴ after the Cocoanut Grove Night Club fire in Boston, which killed 491 people and seriously injured many more. Only a few of the 114 victims taken to the Massachusetts General Hospital had extensive surface burns. Of the 75 who were dead on arrival, the cause appeared to be anoxia caused by inhaling irritant gases and by carbon monoxide poisoning. Only 3 of the 39 who survived long enough to receive

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