

# **Myelomeningocele: An Active Approach<sup>1</sup>**

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Children born with myelomeningocele present possibly the most rapidly increasing population needing orthoses. This rapid increase is attributed to improved life-saving methods introduced by neurosurgeons and urologists. At the same time there is an increased interest in maintenance and restoration of musculoskeletal function by orthopaedic surgeons.

All too often, however, the external support fits poorly, and procurement procedures delay delivery and repairs. Methods for decreasing requirements for support often are not used, and the patient is therefore, committed to prolonged, unnecessary bracing. Bulky supports that are difficult to apply may be

discarded by older, heavier patients, thus negating previous efforts.

Our activities have been directed to the coordination of our orthotic and orthopaedic energies in the most aggressive, yet economical, approach possible for the care of these children. As a result, many hitherto heavily braced patients are ambulating with decreased support or no support at all; other, presumably, unbraceable children are able to use external supports; the overall drain on economic and physical resources of others seems to have been reduced; and a general optimistic attitude prevails in the families of our myelomeningocele patients.

## **THE PROBLEM OF MYELOMENINGOCELE**

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Myelomeningocele (*spina bifida cystica*, "open spine") afflicts approximately one of each 1,000 newborn babies. Its incidence is the

highest in rural, white families, and is as much as 15 times higher in families with another myelomeningocele child.

### Nature of the Disease

The lesions are located anywhere on the spine from the head to the sacrum, but most are in the lumbar spine. Neurologic deficit may range from minimal to complete distally from the level of the lesion. It is not uncommon for significant areas of neurological "escape" to occur, i.e., a child with a lumbar lesion whose one leg is normal while the opposite is severely involved, or the child with a lumbosacral lesion where the inverters of one foot are active and the dorsiflexors and everters of the other are active. In the case of high lesions there is often preservation of reflex function distally, so that the patient may have spasticity as well as paraplegia. (Occasionally patients resist passive motion quite consistently, and we have mistakenly "underbraced" them thinking that such a function was useful.)

### Early Management

Significant progress has been made in two areas of prime neurosurgical interest: closure of the defect and management of hydrocephalus. Early closure of the neural defect reduces mortality, morbidity, and cost of nursing care. Its effect on motor function is not completely known. New techniques and improved devices have decreased the hydrocephalus problem to proportions that pose no significant threat to head control or to ambulation, at least from the point of view of size.

A number of urologic problems are common to myelomeningocele patients. The threat to life of hydronephrosis and pyelonephritis has been reduced considerably by urinary diversion procedures. Incontinence becomes more of a problem with increasing age. External infection of the genitalia, cystitis, and peroneal decubiti are recurrent problems in many of these children.

Strabismus, or squint, is present to some degree in most myelomeningocele patients, though not necessarily associated with vision difficulties. Management is best handled by an ophthalmologist familiar with myelomeningocele patients.

### Problems of the Parents

The physical complications of the myelomeningocele patient are just part of the problems faced by these children and their families. Divorce in families with a handicapped child is not uncommon. Proper counseling about the unknown cause of the defect and its incidence, we believe, could prevent some of these divorces.

Aside from conflict between the parents, myelomeningocele can cause other anxieties in the family. Early counseling and assistance by the visiting public health nurse can minimize the impact of urinary and rectal incontinence, sores, contractures, acute complications such as shunt dysfunction and urinary tract infection, and financial drain on the family.

### Problems of Finance

The financial burden of this disease is onerous. Few of our patients could manage without the assistance of the Bureau of Crippled Children

Services. Moreover, the Bureau is concerned with the general welfare of the child, often in contrast to the compartmentalized views of medical specialists.

The logistics of transportation and time probably present the most difficult paramedical problem for families of these children. There are no funds to reimburse parents for loss of income, and few, if any, monies to pay the often sizable transportation bills for the typical, rural, low-income family affected.

### **Role of the Pediatrician**

Myelomeningocele patients have the same pediatric problems as normal children, complicated by such medical problems as seizures, mental retardation, incontinence, and constipation. A pediatrician is the logical person to supervise and coordinate management of these children, preferably through a special clinic. Since patients often live far from the clinic, routine preventative and medical emergency needs may be handled most efficiently by a well informed medical person in the patient's neighborhood.

In a specialized clinic only a small number of patients can be handled effectively in a single day by a team of experts. Because of the large numbers (15 to 25) that attend our clinic at each session, only the neurologic, orthopaedic, orthotic, and acute social or medical needs can be handled in one session, and as a rule, general pediatric care is provided by practitioners near the home. Other problems, such as routine urologic and ophthalmologic problems, often are approached at another time. The ex-

treme complexity of the problems of myelomeningocele patients demands the highest degree of cooperation and coordination of efforts of all involved in the care of these children and their families.

### **GENERAL ORTHOPAEDIC CARE**

Fewer and fewer children born with myelomeningocele die at birth or soon thereafter. They live and develop crippling and life-threatening musculoskeletal deformities, and therefore it is obligatory to include the orthopaedic surgeon in early evaluation and the consequent programming of care.

### **Evaluation**

Generally speaking, orthopaedic problems are the most common ones for older children, but attention to these problems must begin at birth. In our institution, the orthopaedic surgeon is consulted as soon as the new-born is admitted for primary treatment. At this time, a general assessment of deficits and potential problems is made. Most important is a prediction of potential problems and a general decision about staging of procedures. Few operative procedures can be carried out early.

Our surgical approach is based on the following major principles: 1) removal of deforming forces, 2) alignment of joints in a functional position, 3) replacement of freed tendons to a place where they are most likely to be effective, 4) provision of immediate and continuous external support as deemed necessary, and 5) reduction of the amount of external support as much as possible by "internal bracing",

e.g., tendon transfer. When all procedures cannot be performed at the same time, the following general rule of thumb has been developed for priority of procedures that are obviously needed: 1) foot reconstruction is completed by six months of age, 2) hip surgery is completed by 14 months of age, and 3) spine procedures are completed by three years of age.

### Management Programming

The expense of surgery can be appreciable. We have controlled this by two means. First, as many systems as can be serviced at one time are cared for. As an example, intravenous pyelograms, hearing tests, strabismus evaluation, tonsillectomy, herniorrhaphy, and circumcision are routinely carried out during the same admission. Second, more than one surgical procedure often is performed at one time. Most important, a given joint is completely repaired at one sitting. For example, the procedures for most dislocated hips consists of an iliopsoas transplant, an open reduction of the hip with modified arthroplasty, and a derotational femoral osteotomy to correct the inevitable anteversion, all carried out together. Both sides may be corrected at the same time, and when technically possible, knee and foot procedures are combined with hip surgery. In some cases as many as 12 procedures, when as many as 16 incisions are necessary, have been performed at one sitting. Such extensive operations may last as long as four hours, but with present-day anesthesia and aseptic techniques complications have been held to a minimum.

Procedures are staged so that the elapsed time in casts is minimal. For example, a spine reconstruction, requiring 16 weeks immobilization, may be planned along with hip reconstruction, requiring 6 weeks immobilization. The hip surgery, therefore, is performed 10 weeks after the spine procedure so that all trauma procedures will mature simultaneously.

### Morbidity

Morbidity from surgery is minimal in anesthetic patients. We have yet to reach a limit of "tolerable" surgery, even though we are constantly made aware of the notion that a certain amount of surgery is all that a child can tolerate.

Morbidity is minimized by three means other than the reduction in the number of operative sessions. Hospitalization time is reduced as much as possible. Isolated knee or foot procedures are done on an out-patient basis. Patients with single or multiple major-extremity procedures are kept in the hospital only two or three days until fever is subsiding and they can be cared for by a well-informed parent. Major spine reconstruction is handled during a four- or five-day hospitalization period. It is our experience that routine procedures such as bladder credeing, rectal stimulation, cleansing, and routine care of the cast can be handled by the parents as well as by nursing personnel, and at a cost considerably lower.

### Orthotics

External supports are applied early in life. In addition to the usual benefits, this approach enables us

to manage any fractures that might occur without the need for hospitalization. The use of orthoses also prevents recurrence of contractures.

### NEONATAL CARE

All newborn babies with myelomeningocele are seen at or near the time of admission. Often an evaluation of the functional status of pre- and post-myelomeningocele closure can be obtained.

On the basis of our experience in the examination of these babies, we are convinced that 1) accurate assessment of individual muscles is impossible, 2) little change in gross function occurs with closure, 3) orthopaedic care, which consists primarily of exercises during this period, is facilitated by closure, and 4) a reasonably accurate evaluation of functional potential, particularly in terms of procedures which will be necessary, can be made at this time.

Neonatal cast correction of foot deformities has been abandoned. The feet that need correction most usually are rigid enough to resist correction and sufficiently anesthetic to develop pressure sores as well. Early operative correction is done only when frequent postoperative followup is possible, and when parents are interested and capable of carrying out an active stretching program. Otherwise, feet are corrected surgically at a time when adequate external support can be provided by shoes and orthoses.

We feel also that there is no place for closed treatment of dislocated hips in the neonate with myelomeningocele, because this type of dislocation will recur when the hips are straightened. Furthermore, in paralyzed or partially paralyzed patients,

immobilization in a nonfunctional position is often followed by irreversible contractures in that position. In addition, external rotation, abduction and flexion contractures are the most difficult hip contractures to stretch, and this is the deformity produced by a contracted iliopsoas.

### INFANT CARE

Most major reconstruction, in our opinion, should be completed before the child is ready to walk. For a nearly normal child this is within the first 18 months of life. Higher paralysis and more serious central nervous-system deficits extend the time before the child will want to walk. Indeed, certain children have no potential for walking. However, we do not accept significant contractures in any but the most completely decerebrate patients. Almost all have sufficient control of the trunk to sit, and most parents want to put shoes on their children.

Standing is often a very acceptable goal for a youngster, one which is most appreciated by the patient and parents. We often set this goal for patients under one year of age who have been given braces after surgery for severe instability of the spine or for rigid dislocation of the hips, knees, or feet.

### SALVAGE PROCEDURES

We have been successful in preventing severe contractures in post-operative patients who wear their braces conscientiously. When braces are not worn, or when patients have not been relieved of contractures early in life, it is occasionally necessary to resort to such salvage procedures as joint resection, massive

release of contractures, particularly about the hip, resection of bones (e.g., talectomy), or osteoclasis, ostectomy, and arthrodesis to obtain functional alignment.

Results with such procedures are generally satisfactory, and often are appreciated more than the more prophylactic procedures done at an earlier stage. Seldom, however, can the patients be made as functional by late or secondary surgery as by early or primary definitive surgery.

## SUMMARY OF EXPERIENCE

From 1968 through 1971, 497 procedures were performed on 133 patients during 166 operative sessions that averaged two hours each.

By region there were:

*40 spine procedures*, primarily spine osteotomies with resection of deformed segments.

*211 hip procedures*, primarily iliopsoas transfers, open reduction, and proximal femoral osteotomies for anteversion.

*207 foot procedures*, primarily releases of contractures, tendon transfers, and Grice procedures.

*39 other procedures*, including hamstring transfers, tibial osteotomies, and others.

There were three operative deaths, all in patients undergoing spine osteotomy. Excessive bleeding was the cause. There were no major wound infections, but several foot wounds were delayed in healing.

The major complications consisted of failure to achieve the goals of surgery. Most important, it was discovered that femoral anteversion was always present when hips were dislocated, and therefore, the hip

had to be reduced in internal rotation. A corrective osteotomy is also essential, because external supports cannot be applied effectively unless knees and feet face forward. The osteotomy is performed at the same time as the original surgical attack, and the small proximal fragment is held in the reduced position by an indwelling Kirshner wire until time for fitting of the orthosis.

The other major complication was recurrence of contracture, especially in the foot. Unless external support is applied immediately after surgery, and parents maintain a reasonable stretching program, it is not uncommon to have nearly complete recurrence within weeks.

Secondary and tertiary procedures are always more and more difficult. Occasionally, salvage procedures such as talectomy and joint fusion were necessary as a last resort.

## ORTHOTICS

The key to success of a surgical program for treatment of the myelomeningocele patient is close cooperation between the orthopaedic surgeon and the orthotist. Needless to say, a large number of trials and errors has accompanied the evolution of our external support program. Many problems remain. These include the need for smaller, lighter, interchangeable components; the need for materials which resist excreta, and wear better; the need for supports which can be lengthened and enlarged in part rather than in toto.

Three major hurdles for external support programs have been cleared, however. First, through close coop-

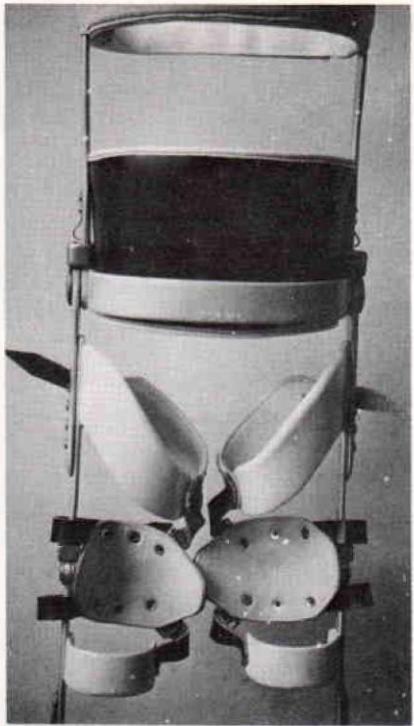


Fig. 1

Finished orthosis. A plastisol is used to cover the thigh and calf bands.

eration, prescription writing has been simplified. Special orders are given only when modifications are desired, but this is not often because our basic design incorporates smaller compact parts and because of the policy of treatment at an early age.

Some of the big changes made were in materials for covering the pads and cuffs, and in the fitting method. A nonabsorbent material that could be applied to the cuffs and pads was most difficult to find. In the pelvis and buttocks area we use Naugahyde instead of horse- or cowhide. The thigh and calf bands are covered with a plastisol and the anterior parts of the cuffs can be

covered with either a plastisol or Naugahyde covered leather (Fig. 1).

Experience in fitting the orthoses prompted us to change our initial design somewhat. The original objectives were to help prevent recurrence of contractures and to control rotation. The area that had to be changed drastically was the hip-thigh complex. We were concerned primarily about anteroposterior control, and to a great extent overlooked the need for medial and/or lateral control. When iliopsoas transfers and proximal femoral osteotomies were carried out, it became apparent that a better lateral control was absolutely necessary to provide the best conditions for healing. We then redesigned the orthoses to provide a fit more snugly laterally at the hip-joint level, and more importantly, a tighter lateral fit on the shaft of the femur at the thigh-band level (Fig. 2). To provide an even,

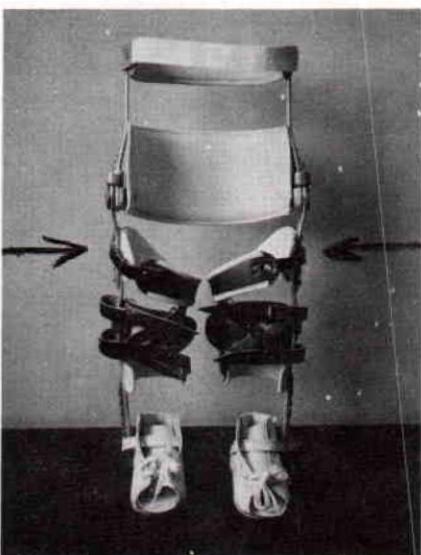


Fig. 2

View of a finished orthosis showing the snug fit along the lateral aspect of the thighs.

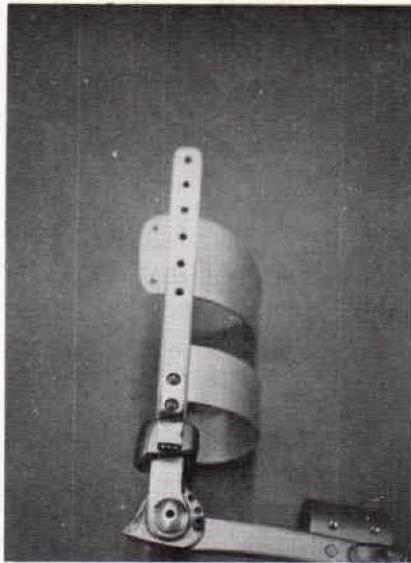


Fig. 3

View of thigh band showing the extension beyond the lateral upright to provide optimum stability.

firm force, the thigh band was redesigned so that it extends forward of the lateral upright by a distance equal to approximately the width of the femur (Fig. 3).

To help with the problems imposed by diapers the thigh band was designed to be much lower on the medial side than in conventional orthoses (Fig. 4).

We have found also that most orthoses for these children needed only lateral uprights for the legs (Fig. 5).

One big mistake that we made was in the method of attaching the ankle joint to the shoe. To make the orthoses easier to fabricate we used straight uprights with no joints from the calf area to the shoe. Continuous breakage and malfunction occurred. We reverted to the use of ankle joints, permitting a little anterior and/or posterior mo-

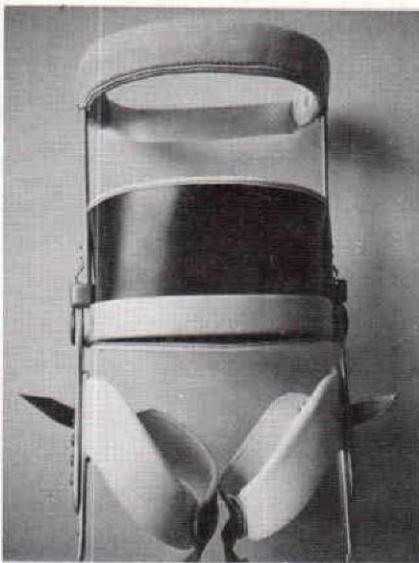


Fig. 4

Close-up view of thigh bands with lower medial sides to help with problems imposed by diapers.

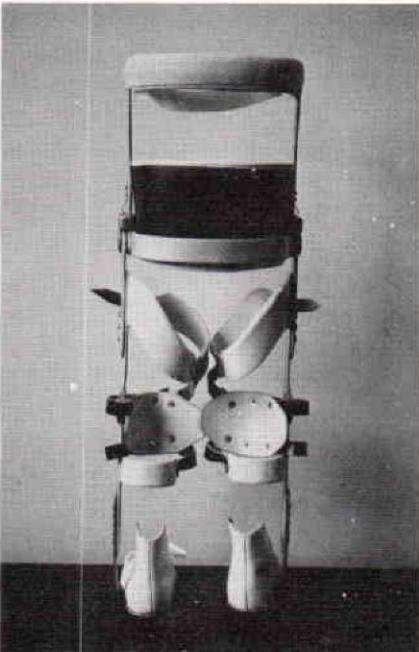


Fig. 5

Posterior view of the orthosis generally used in treatment of the child with myelomeningocele.

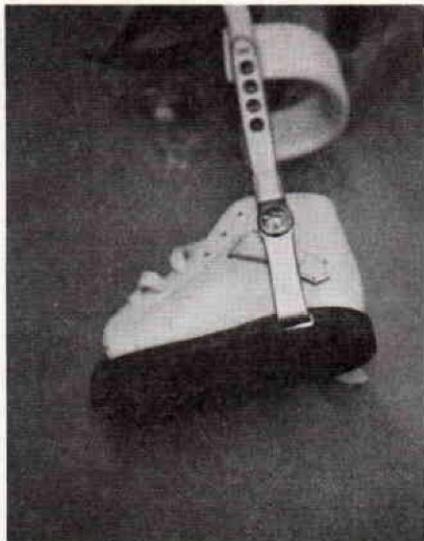


Fig. 6

Close-up view of the ankle joint and shoe attachment.

tion, and the breakage virtually ceased while function was excellent. Our big problem was finding a joint and shoe attachment small enough. These joints (Fig. 6) are now commercially available.<sup>4</sup>

Although many problems arise when a thoracic extension is added above the hip, the need for this is quite evident in myelomeningocele and cerebral palsy patients. When the earlier management procedures were employed, the thoracic extension required a thoracic band with chest straps, a lateral corrective strap, and a full abdominal front, in an attempt to correct lordosis and scoliosis. With the new and more advanced surgical techniques and the recognition of "surgically correcting" and "orthotically holding" the design of the thoracic extension has been simplified. We now use lateral extensions with only a thor-

acic band and chest strap. No pelvic or belly strap is necessary (Fig. 7). This is very handy and can be a conversion from a regular pelvic band and pad when the patient has to have an "ileostomy" or "Bricker Procedure" for kidney problems. Many times just a full chest strap of leather or cloth webbing is used with the lateral extension (Fig. 8). No band is necessary and the fitting of clothing is much easier. The important thing about the thoracic extension is the additional lever arm provided above the hip joint to approximate the lever arm below the hip joint.

The aluminum alloy parts that are commercially available result in a very light orthosis. Research is

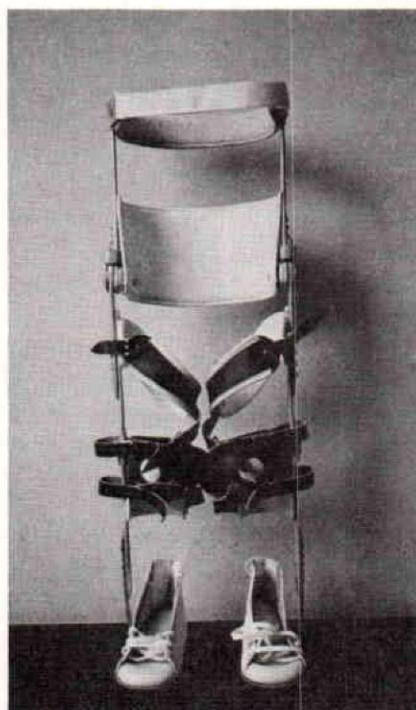


Fig. 7

Anterior view of the orthosis generally used in treatment of the child with myelomeningocele.

<sup>4</sup> Finnieston Laboratories, 1901 N.W. 17th Avenue, Miami, Florida 33125.

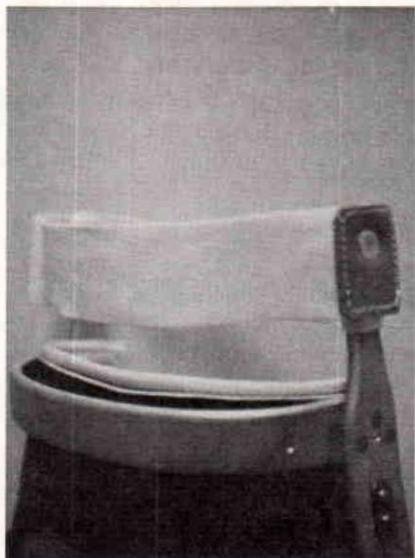


Fig. 8

Close-up view of lateral extension and simple chest strap of cotton webbing.

being carried out using plastics that we hope will help in further weight reduction.

Second, time between surgery and application of orthoses has been reduced. Patients are routinely measured just prior to surgery or during a postoperative cast change. Orthoses are then made available when the casts are removed. Unless the transition from casts to braces is immediate, the incidence of fractures and loss of correction rises at an alarming rate. This has been possible because we have utilized the New York University lower-extremity measurement and layout technique and when this procedure is followed no or very little adjustments are required.

And, finally, through the close cooperation of third party payers, especially the Bureau of Crippled Children Services, and the orthotists, supports are available exactly

when they are needed. A system has been developed for fitting in the offices of the surgeons so that adjustments can be made at the time of primary fitting.

### Overall Concept

Our general philosophy of bracing is as follows:

Early bracing enables children to stand and encourages attempts at ambulation. Early bracing also helps prevent fractures. Those uncomplicated fractures which do occur are treated on an outpatient basis. This results in significant savings of time and money. The major disadvantage to early bracing is the need for frequent revisions of the support system. This is overcome, at least in part, by inspection at least every three months. If revisions are needed, they are made the same day.

Orthoses are applied only to support weak parts or to maintain alignment. Treatment of contractures is, in our view, a surgical problem. It is our aim to have every support system applied by one person without any force. A minimum of corrective straps should be employed in order to minimize the complexity of the support system and thus encourage its use.

Bracing to permit standing is a legitimate goal. We believe that it enhances body tone and stimulates interest in the child's environment. Parents become more enthusiastic, and more activity occurs in the physical therapy area at home. Patients who have severely compromised trunk or head control cannot be braced effectively, and we delay application of external support in-

definitely unless it is required to supplement release of unmanageable contractures.

Every attempt should be made to remove external supports as soon as practical. Hip reconstruction has freed at least five patients from control braces. A few others have been able to abandon "long-leg" braces after hamstring transfers. Continuous efforts are being made to devise

means to eliminate the need for external supports. The extreme value of rendering patients free of braces is two-fold. First, costs of care are reduced. Second, brace wearing becomes more and more difficult as children grow larger and/or become required to apply them without assistance. Few adults continue to wear control braces for these reasons.

#### ILLUSTRATIVE CASE REPORTS

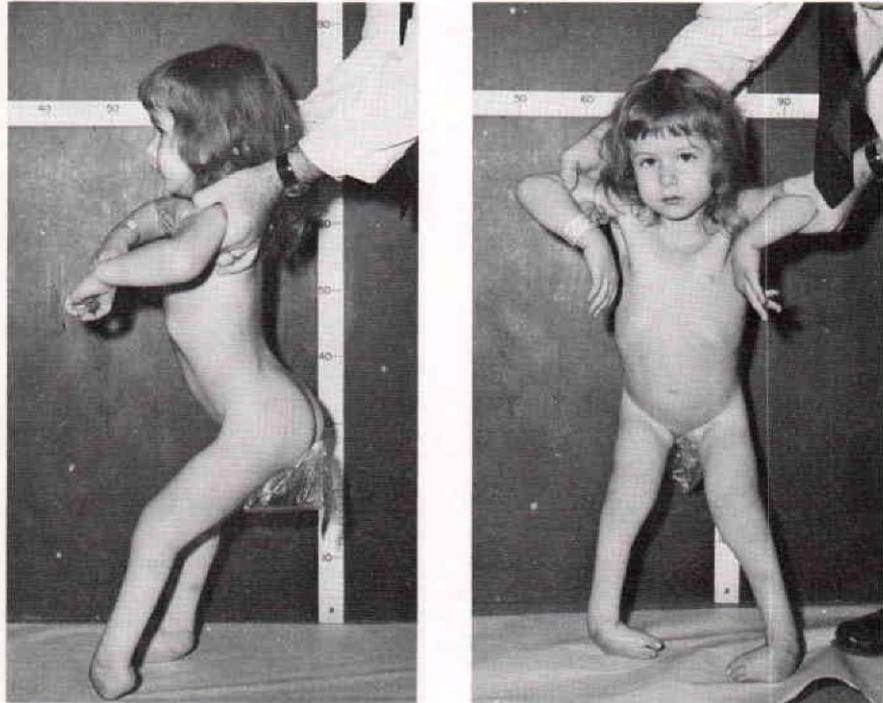


Fig. 9  
Lateral and anterior views of Case 1.

Case 1: R.S., a six-year-old girl, was first seen at the age of three, at which time she had dislocated,

contracted hips and feet (Fig. 9). She had been under nonoperative treatment for her entire three years

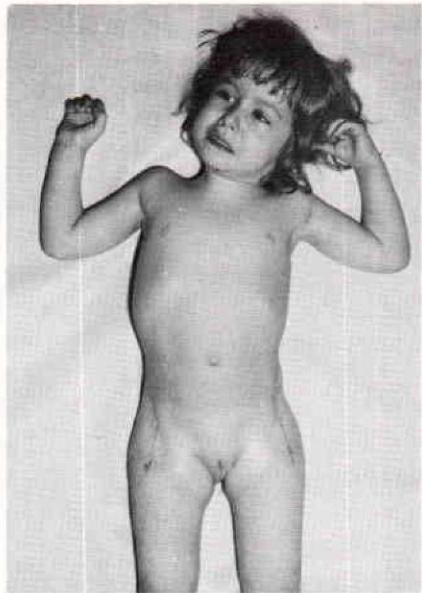


Fig. 10  
Two views of Case 1 after treatment.

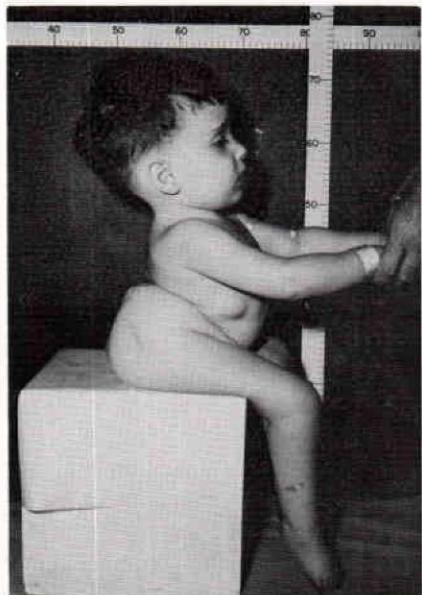


Fig. 11  
Lateral view of Case 2.

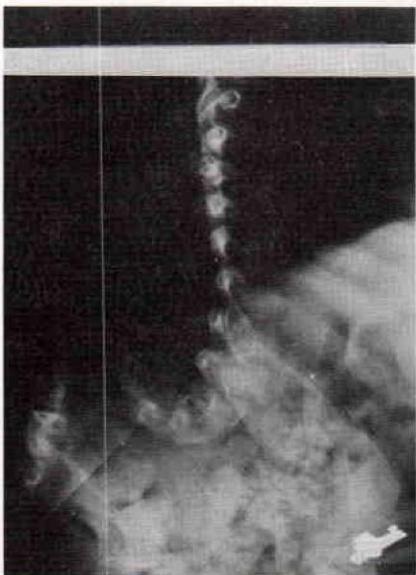


Fig. 12  
X-ray of spine of Case 2 before operation.

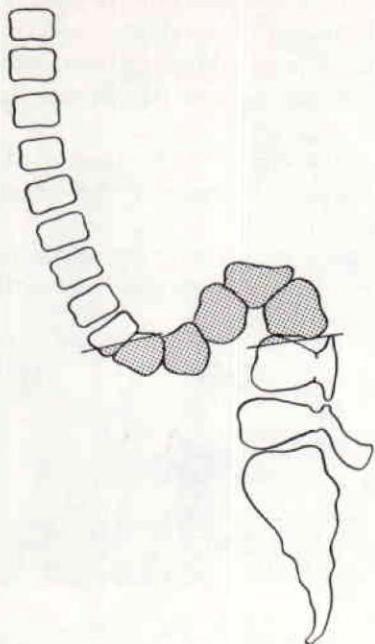


Fig. 13

Schematic view of spine resection carried out on Case 2.

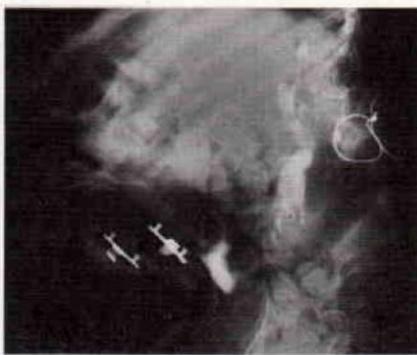


Fig. 14

X-ray of Case 2 after spine resection and fusion.

of life, with approximately 150 days having been spent in the hospital. The problem was unchanged by this therapy.

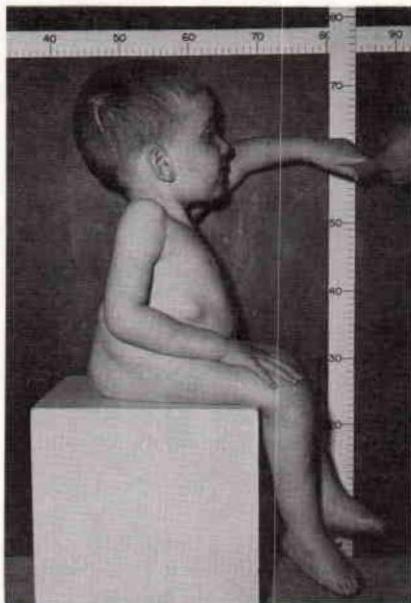


Fig. 15

Lateral view of Case 2 after treatment.

Bilateral iliopsoas transfers, acetabuloplasty, open reduction of hips, and femoral osteotomy were combined with bilateral foot releases. Control braces were applied one month later and ambulation began immediately (Fig. 10). After one year, braces were eliminated and the child now walks with crutches. Small knee-flexion contractures are present, but acceptable.

**Case 2:** T.K., a seven-year-old boy was seen at the age of four because he was unable to wear control braces. He had a large kyphosis over which the skin became ulcerated (Figs. 11 and 12). He could not lie on his back and ambulation, though desired, was not possible. An additional problem was impingement of his ileostomy bag on his thigh when he sat. The bag often

fell off. Spine resection (Fig. 13) and fusion resulted in a stable spine (Fig. 14) and the patient has been ambulatory with crutches and braces since five months after the operation. The problem concerning the iliosotomy bag was solved also (Fig. 15).

### SUMMARY

A team of interested specialists can provide effective care for pa-

tients with multiple birth defects.

An active surgical approach to orthopaedic problems can reduce cost and increase the average functional level.

Essential to the approach is close cooperation between the surgeon and orthotist.

New means must be sought continually to reduce costs and further increase function.