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Rehabilitation problems in spina bifida

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Spina bifida covers a wide variety of abnormalities of development of the spinal cord and central nervous system. The lesion may vary from the occult form in which there is only abnormality of the spinous process or laminae to extensive defects with severe developmental abnormality of the spinal cord. The most important lesions are those constituting spina bifida cystica and of these there are three types.

Meningocele, in which there is a skin-covered swelling but in which there is no neurological deficit and no evidence of any neural tissue within the sac, constitutes only about 7% of all lesions of spina bifida cystica. If the skin-covered swelling is operated upon within the first 2 or 3 days of life, there should be no neurological abnormality and rehabilitation of the child is unnecessary. If, however, the sac is allowed to rupture or becomes infected, secondary involvement of the nervous system can sometimes lead to neurological deficit and secondary orthopaedic defects.

Spina bifida with lipoma occurs more commonly, in about 14% of patients with spina bifida cystica. It may be noticed at birth by the presence of a swelling in the lumbar region or the presence of neurological deformity in the lower limbs, sometimes as a variety of talipes equinovarus. Sometimes the child presents as a problem of enuresis; the child fails to gain control of the bladder at the end of the first year of life and it is then discovered that there is a lesion in the lumbosacral spine. More usually, the swelling in the lumbar region is noticed in the first 3 months of life or later. In almost all these patients there is a liability to the development of progressive paralysis of the lower limbs and bladder during the first 5 years of life. Ideally, the lesion should be operated on to release constrictive or adherent neural tissues and to remove part, but no all, of the lipomatous mass.

Myelomeningocele is the commonest variety of spina bifida cystica. This is the most serious abnormality in which the neural tissues are abnormal and present on the surface of the body as an open plaque of nervous tissue. The defect in the spine and neural tissues may be very small, when it usually presents in the sacral or thoracic region and involves only one or two bony segments, or it may extend as a complete lesion from the mid-thoracic to

the sacral spine. A high proportion of patients born with myelomeningocele show evidence of partial or complete paralysis of the lower limbs and bladder at birth. The paralysis involves both sensory and motor nerves and may be of lower motor neurone or upper motor neurone type or a combination of both. The lower limbs may be undeformed or may show a variety of deformities including dislocation of the hip, recurvatum of the knee, talipes equinovarus, talipes calcaneus, talipes calcaneovarus, convex pes valgus or pes cavus with claw toes. These deformities are *not* congenital deformities of the ordinary kind but are paralytic deformities arising in utero. Because of the paralytic nature of the deformities and the presence of sensory loss, ordinary methods of orthopaedic treatment of these deformities, such as might be appropriate in congenital dislocation of the hip or talipes equinovarus, should not and cannot be applied.

More than 90% of patients with myelomeningocele have partial or complete bladder paralysis. This also complicates their management. Sometimes paralytic hydronephrosis is also present at birth. In others, urinary infection develops rapidly and may give rise to serious secondary consequences in childhood.

In 80% of patients with myelomeningocele there is an associated hydrocephalus, and in 50% the hydrocephalus is likely to become severe and require surgical treatment.

Although this paper will be primarily concerned with the orthopaedic rehabilitation of the lower limbs, treatment must always be considered in relation to the whole child and the management of his multiple disabilities. The management must be undertaken by a team of neurosurgeon, orthopaedic surgeon, urological surgeon, paediatrician, physiotherapist, social worker and orthotist.

Initial assessment

The rehabilitation of a child with spina bifida starts on the first day of life. Soon after birth, the baby may be moving his lower limbs quite vigorously. If the lesion is left untreated the surface becomes dry and later infected and the plaque is raised from the surface of the body by the collection of cerebrospinal fluid beneath it, causing traction lesion on the spinal roots. Within a few hours of birth, movements in the lower limbs diminish so that by the third or fourth day there may be complete paralysis of the limbs which then becomes permanent. To avoid these consequences, the spinal lesion should be treated by urgent operation, preferably within the first 12 hours of life.

The child needs to be transferred to a centre where neurosurgical or paediatric surgical facilities are available for a neonatal operation. Care needs to be taken to avoid excessive cooling because of the lack of normal thermal responses in the newborn period. The spinal lesion needs to be covered with a non-adherent saline dressing and the baby kept in an incubator to maintain his body temperature. Blood is cross-matched for operation.

A general and orthopaedic assessment needs to be made as soon as the child is fit for examination. General examination needs to be made for any other abnormalities such as congenital cardiac or renal lesions. The head should be examined for the presence of hydrocephalus; significant hydrocephalus can often be recognised by the presence of an open lambdoid suture. The spinal lesion is assessed as to size and extent and the presence of any spinal deformity, such as spinal kyphosis, is recognised and radiographs taken of the spine.

The lower limbs are examined to determine the presence of any deformity at each joint with particular attention to the range of abduction and the presence of any fixed flexion at the hip, recurvatum or fixed flexion deformity of the knee or any talipes deformity of the feet.

Voluntary activity in each muscle group can be assessed by stimulating the child to become irritable; if he is crying and moving his upper limbs, any muscle activity that is present in the lower limbs will usually be obvious. Activity in the lower limb muscles is either recorded as present or not present and the presence should be charted in order of root innervation. Examination of voluntary movement should always be supplemented by percutaneous faradic stimulation of nerves and muscles in the lower limbs. A child who may appear to have complete clinical paralysis of the lower limbs will often show a good response to faradic stimulation – indicating that any paralysis is temporary and may recover once the spinal lesion has been treated.

A number of patterns of paralysis and deformity are easily recognisable. In a child who has innervation down to the third or fourth lumbar segment with paralysis below this level, he presents at birth with flexed, adducted, dislocated hips, recurvatum of the knee and varus deformity of the feet. The active muscles that are present include all the hip flexor muscles, all the hip adductors, quadriceps and tibialis anterior; all other muscles are paralysed. The radiograph shows bilateral dislocation of the hips.

A child innervated down to the fifth lumbar segment with paralysis below this shows some flexion deformity at birth of the hips and limitation of abduction with subluxation. There is usually no significant deformity of the knee. The feet show calcaneus or calcaneovarus deformity. The muscles that are active include all the hip flexors and adductors, some action in the hip abductors with paralysis of the hip extensors, normal quadriceps and some action in the semitendinosus and semimembranosus and activity in most of the dorsiflexors and evertors of the foot. The radiograph shows subluxation of the hips.

In a child innervated down to the first sacral segment but with paralysis below this, there is no deformity of the hip or knee. The feet show clinical convex pes valgus deformity and the radiograph shows a vertical talus. Most muscles in the lower limb are active except for the biceps femoris, flexor digitorum longus and the intrinsic muscles of the feet. There is weakness of the triceps surae and peronaeus longus.

If there is innervation down to the second sacral segment, the only de-

formity that presents is a mild clawing of the toes associated with relative weakness of the intrinsic muscles of the feet.

Initial treatment

Unless the child is extremely severely affected and unlikely to live, operation on the spinal lesion between the sixth and twelfth hour of life gives the best chance of diminishing any secondary paralysis of the lower limb muscles. The aim of the operation is to provide adequate covering of the spinal cord without damage to it. The central plaque of neural tissues, although an abnormal shape, contains active nerve cells and this can be demonstrated by electrical stimulation of the plaque area which causes movements in the muscles of the lower limbs. This area is preserved and the membrane between it and the skin is removed. A layer of dura is dissected up from the wall of the spinal canal and sutured to reform the dural tube. The closure of the dura should not be too tight or it may lead to secondary pressure on the spinal cord in succeeding weeks of life. Sometimes a further layer of covering by fascia can be obtained but this is not always possible. Skin closure is almost always obtainable by mobilisation of the skin surface of the back and the abdominal wall with vertical closure and occasionally transverse closure. Only in exceptional cases it may be necessary to perform relaxing incisions or Z-flap incision.

If there is severe lumbar kyphosis, skin closure is made extremely difficult if not impossible. Closure of the spinal lesion then requires performance of osteotomy of the spine with excision of vertebral bodies. They are then sutured and maintained in position by Kirschner wires. After the operation, the baby is nursed in an incubator and healing in most patients complete within 7 days.

As soon as the spinal wound is healed, attention needs to be paid to the presence of hydrocephalus. Ventriculography can be performed at the third week of life. If the intraventricular pressure is more than 300 ml or the depth of the cerebral cortex at the vertex is less than 15 mm, ventriculo-atrial or ventriculoperitoneal shunting of the cerebrospinal fluid is needed. In some instances, it may be appropriate to delay a decision about the need for shunting until the child is 3 months or more, but it is important to recognise that hydrocephalus seldom ceases spontaneously and, once a shunt has been inserted, it will need to be observed carefully throughout life and renewed at any time when there is evidence that it is becoming blocked or otherwise ceasing to function. It is not the purpose of this paper to discuss the further management and complications of the mangement of hydrocephalus; suffice it to say that, on average, shunting needs to be revised approximately every 5 years.

Ventriculitis and meningitis are a constant danger in any child with myelomeningocele; it is most likely to develop during the first week of life whether the lesion is operated upon or not, but it may develop at any time later in life even in those who have had a satisfactory ventriculo-atrial shunt.

At the 3rd or 4th week of life attention also needs to be paid to the condition of the urinary tract. Intravenous pyelography to determine the presence of any renal or bladder abnormalities is performed as routine but should be repeated at least once a year after that. Repeated tests of the urine for any evidence of urinary infection need to be made and estimates of renal function by blood urea and non-protein nitrogen tests are needed, especially before any operative treatment for orthopaedic or neurosurgical purposes is made.

Even though there may be obvious deformities in the lower limbs at birth, orthopaedic treatment is seldom of value during the first 12 weeks of life. Even if the child has complete dislocation of the hips, the deformity is so severe that application of splints such as the van Rosen splint is impossible. The deformities of the feet are often severe and fixed and the application of any forced splintage to attempt to correct deformity is extremely liable to result in the development of pressure sores or gangrene of the toes. During the early weeks of life, the main need is for treatment of the spinal lesion and hydrocephalus and the measures needed for these taken priority over any orthopaedic management. Simple passive movements to maintain as much mobility in the limbs as possible can be employed by physiotherapist or taught to parents.

The principles of orthopaedic treatment

The aim in the orthopaedic management of spina bifida is to correct deformity, to maintain correction and to obtain the maximal functional use of the lower limbs. Certain common principles apply in the management of the lower limbs.

1. Assessment. Apart from the assessment at birth, further assessment of the extent of deformity at each joint by clinical and radiological examination needs to be made at the third month and then at 6 monthly intervals, throughout the growing life of the child.

2. Muscle activity. Periodic assessment of muscle activity, especially in relation to any deformity that may be present, should be made at the third and sixth month of life and at 6-monthly intervals after that time. As the baby becomes older, it is possible to define more accurately the presence of true voluntary movement or reflex activity. All varieties of neurological lesion may be present either at lower or upper motor neurone level or a combination of both. Sometimes, the presence of activity in reflexly innervated muscles may only be able to be determined with certainty by electrical stimulation. Electromyography is occasionally useful but is of much less value in clinical application than simple electrical (faradic) stimulation. Sensory testing is difficult to perform in small children and is not reliable until a child is more than 6 or 7 years old. In cases of doubt it is wise to assume that there is sensory loss that corresponds to the level of motor innervation.

3. Correction of deformity. Deformity can sometimes be corrected by careful

application of splintage but it is seldom possible to make complete correction by conservative means and it may at times be very dangerous to attempt to do so by splints, plaster or frames that might be appropriate in the correction of non-paralytic deformities. Deformity should preferably be corrected by operative means, by radical division or elongation of short tendons, muscles and fasciae. It is surprising how much correction of the deformity can be obtained in children aged between 6 months and 2 years by division of soft tissues and tendons alone. If division of soft tissues and tendons fails to give complete correction of the deformity, it may then be appropriate to consider further correction by osteotomy of the upper or lower end of the femur, of the tibia at its shaft or in the supramalleolar region or in the foot at the calcaneus or mid-tarsal bone.

4. Conservative or operative correction of the deformity must be followed by *restoration of balanced muscle action* at each joint. It is useless to correct deformity and to leave unbalanced musculature that would be capable of acting and causing a recurrence of the deformity. The application of splints such as night splints will not prevent the recurrence of deformity and they are very liable to cause pressure ulceration. They should not be used. If muscle activity has been correctly analysed, it should be possible to balance muscle activity by tendon transplantation of stronger muscles to replace those that are paretic or paralysed.

5. Fixation. Wherever possible, splintage or fixation of any kind should be retained for a minimal period. After operation, it is seldom necessary to maintain plaster fixation for longer than 3 to 4 weeks for soft tissue release or tendon transplantation or for longer than 6 weeks following osteotomy. Long periods of fixation are likely to produce secondary deformity and increase the liability to spontaneous fracture when the fixation is removed.

6. Bracing. No child should be encouraged to stand or walk before deformity has been corrected, particularly at the feet. The need for bracing will naturally vary with the extent of paralysis in the child. A child who remains severely paralysed or who has only reflex function in the lower limbs will need to be supplied with a brace extending from the chest to the feet. Such bracing needs to be carefully made and applied and should be used to stabilise the joints over which the patient has inadequate or absent control. It is seldom possible to train a severely paralysed child to walk in extensive splintage until he is at least 3½ years old. Other less paralysed children may need less extensive splintage, especially if adequate tendon transplantation has been performed to allow spontaneous stabilisation of the hips and knees so that only appliances below the knee are required.

Since most spina bifida children have normal upper limbs and a strong trunk, almost all of them can be made capable of walking even though this may be for short distances. Addiction to a wheel-chair should be avoided because of its liability to result in increasing obesity, spinal deformity, porosis of the limb bones with spontaneous fractures and pressure sores in the ischial region.

Subluxation or dislocation of the hip is present at birth or develops during the first 3 years of life in more than half of all children suffering from myelomeningocele. It arises when there is strong action in the hip flexor and adductor muscles with paresis or paralysis of the gluteal, abductor and extensor muscles. The acetabulum is often preserved remarkably well even though the hip is dislocated at birth. The neck of the femur often shows anteversion and valgus deformity but this does not prevent reduction of the dislocation provided that the muscles surrounding the hip joint have been adequately mobilised.

Any adductor deformity needs to be corrected between the 3rd and 12th month of life by radical open adductor tenotomy. Subcutaneous tenotomy is not sufficient to correct muscle length and it is often necessary to divide all the adductor muscles at their attachment to the pelvis with preservation of the obturator vessels and nerves. The aim should be to obtain full abduction of the hips and the hip will then reduce in most instances. Even in a child in whom dislocation is present 3 or 4 years after birth, reduction is often possible provided that adductor release has been performed adequately. Reduction is then maintained by application of plaster or splintage in extension, abduction and medial rotation, but splintage should not be maintained continuously for longer than a month.

The second stage in the procedure is to correct muscle balance by posterolateral transplantation of the iliopsoas muscle. Ideally the muscle should be transplanted 1 or 2 weeks after adductor tenotomy has been performed and the operation can be done at any time after the 6th month of life. The combined iliopsoas tendon is detached from the lesser trochanter by an approach between the femoral vessels and nerve, and mobilised into the pelvis. The whole of the iliacus is detached from the inner surface of the os ilium. A foramen is made just lateral to the sacroiliac joint through which the whole of the iliacus and the iliopsoas tendon is passed into the buttock. The tendon is then re-attached to the posterior aspect of the greater trochanter, the tendon being passed through a bony tunnel to be sutured to the trochanter. The combination of adductor release and posterior iliopsoas transplantation will allow reduction of almost all dislocated hips. In some instances it may be necessary to perform a capsulorrhaphy of the hip joint but it is seldom ever necessary to perform varus osteotomy of the femoral neck. The normal varus angle of the neck of the femur will be restored spontaneously as a result of the action of the iliopsoas on the greater trochanter.

The transplanted iliopsoas will prevent redislocation of the hip. It also helps to stabilise the hip for walking so that, in many instances, it will not be necessary to apply any bracing to the region of the hip to allow walking.

In some children, particularly those who are severely paralysed, the main deformity may be a flexion and lateral rotation deformity of the hip, usually

due to powerful but isolated action of the iliopsoas and sartorius muscles. In these children, the hips tend to lie in flexion, abduction and lateral rotation. The hip does not dislocate but bracing cannot be applied to allow walking because of the extent of the deformity. Correction needs to be made by division or elongation of any tight flexors and, in severe cases, intertrochanteric extension osteotomy. It is often useful to transplant the iliopsoas tendon to the anterior aspect of the greater trochanter so that it may act as a medial rotator.

Knee deformity

Fixed extension or recurvatum of the knee may be present at birth in children who have a normally acting quadriceps in the presence of paralysis of the knee flexor muscles. In many children the range of flexion gradually returns with the help of simple passive movements but when the deformity is severe, the sartorius and gracilis muscles may be found to be secondarily displaced anterior to the knee joint and they become perverted extensors of the knee. When this situation is present, elongation of the quadriceps may be needed.

Flexion deformity of the knee may arise in children who develop spasticity in the lower limbs or in children with extensive paralysis who have been allowed to sit for long periods with the knee in the flexed position. If there is complete paralysis of all muscles acting on the knee, the division of all tight posterior tendons may be sufficient to correct deformity. If the hamstring muscles are active, one or more of them may be transferred to the posterior aspect of the lower third of the end of the femur.

If soft tissue division fails to allow correction of the deformity, supracondylar extension osteotomy of the lower end of the femur may be needed.

Foot deformity

The most common foot deformities are equinovarus, calcaneus or convex pes valgus. Varus deformity is almost always associated with activity in the tibialis muscles with paralysis of other muscles below the knee. In some patients there may be spastic activity in the triceps surae which aggravates the deformity further. Correction can seldom be achieved by conservative means. It can be achieved with much more safety by operative measures sometime between the first and second year of life. Through a posteromedial incision, all the tight structures on the posterior and medial aspects of the ankle and on the medial and plantar aspects of the foot are divided or elongated. The aim should be to obtain a completely plantigrade correction of the foot and there should be no sparing of ligaments or joint capsules to obtain this. If pre-operative evaluation of muscle activity has shown a strong tibialis anterior muscle, it is usually appropriate to transfer the tendon laterally to the dorsum of the foot in the region of the cuboid bone. If there is a predominance of calcaneus deformity, it may be transplanted

through the interosseous membrane to the tendo calcaneus. If soft tissue division fails to give complete correction of deformity, osteotomy of the calcaneus, partial or total excision of the cuboid, metatarsal osteotomy or excision of the talus may be needed.

Calcaneus deformity usually presents in children with strong activity in the dorsiflexors and paresis or paralysis of the plantarflexor muscles. Correction of the deformity by elongation of the tendons on the dorsum of the foot and ankle with transplantation of one or two muscles through the interosseous membrane to the tendo calcaneus will usually give good correction with balanced action at the ankle joint.

Convex pes valgus deformity with vertical talus requires special attention. There is usually good activity in most muscles of the foot except the intrinsic muscles. The navicular bone is dislocated on to the dorsal aspect of the talus and there is calcaneovalgus deformity of the forefoot and equinovalgus deformity of the hindfoot. Correction always needs to be made by operative means. Through a dorsomedial incision, the tibialis anterior is detached from its insertion and attached to the neck of the talus. All the extensor tendons of the toes are elongated. The dislocation of the talonavicular joint is reduced and the position held with a Kirschner wire. Through a second posterolateral incision, the peroneus brevis tendon is divided from its insertion. The talocalcaneal joint is opened on the lateral side and any ligamentous tightness released to allow correction of valgus deformity. If there is then still some equinus deformity, the incision is continued posteriorly to allow elongation of the tendo calcaneus. The final step in the operation is the transfer of the peroneus brevis tendon behind the ankle joint to the navicular at the point of insertion of the tibialis posterior tendon. This complex procedure will result in full correction of deformity and balanced muscle activity at the hindfoot and forefoot.

Cavus deformity of the foot with clawing of the toes is present in those children with almost normal innervation in the lower limbs. If left uncorrected, pressure ulceration is likely to develop over the dorsal aspect of the toes and over the plantar aspect of the metatarsal heads. The deformity can be corrected easily by transplantation of the long toe flexors in each toe to the extensor surface of the toe and by tenodesis of the flexor hallucis longus to the proximal phalanx of the great toe.

Late deformities

In a child who is able to receive orthopaedic attention within the first 6 months of life, it should be possible to obtain correction of all deformity by the time he is 2 or 3 years old. If deformity has not been corrected, it is still possible to make a satisfactory correction of deformity on the same principles as those already described but there is likely to be some increasing need for osteotomies to maintain complete correction. Whatever degree of paralysis there is in the lower limbs, it should be possible to correct deformity

and to maintain correction so that bracing can be fitted and walking made possible. Since the deformities are frequently present at all joints in the lower limbs, multiple operations may be needed. The average number of orthopaedic procedures required in the lower limb in most spina bifida children is likely to be between 6 and 8.

Spine deformity

Mention has already been made of kyphosis of the spine that may be present at birth and which requires correction by vertebral osteotomy on the first day of life. In some children, increasing kyphotic deformity of the lumbar spine requires that correction be obtained in an older child. Increasing deformity leading to inability to apply bracing or the recurrent development of pressure ulceration over the prominent spine requires correction by osteotomy of the spine usually with excision of two vertebrae and fixation by staples.

Lordosis and lordoscoliosis may develop in a younger child but, more often, severe deformity does not start to develop until the child is 11 or 12 years of age. If uncorrected, the deformity is likely to become severe. It may be possible to correct deformity by posterior soft tissue division and the application of Harrington rod fixation and spine fusion but, if lordosis is severe, posterior correction cannot be made. In this situation, anterior retroperitoneal correction of lordoscoliosis with excision of intervertebral discs and adjoining portions of the vertebral bodies will give good correction and the Dwyer method of fixation by staples and cable has given excellent results up to the present time.

Education in walking

Once all deformities have been corrected, it should be possible to train a child to walk independently. The only exception is where there has been severe and uncorrected hydrocephalus or severe uncorrected spinal deformity. Even if there is no muscle activity in either lower limb, a spinal brace with double long calipers can be applied, provided that lower limb deformities have been corrected completely. The calipers are hinged on to the spinal support so as to allow some hip flexion and abduction so that the child may be able to hitch up his pelvis and to throw his leg forward at each step.

If paralysis in the lower limbs is partial, less extensive bracing may be needed and, if posterior iliopsoas transplantation has been performed at the hip and there is adequate quadriceps action, the only bracing that will be needed is likely to be below the knee. Footwear needs to be carefully made to avoid any excessive pressure at any point on the skin and the lacing of any shoes should be made so that they can be opened at the toe. The parents, and later the child himself, should be taught to look for any early evidence of pressure that may be demonstrated by the development of red-

ness of the skin. If any pressure sore threatens to develop, it must be treated by complete avoidance of any further pressure or bearing of weight until the sore has healed.

Spontaneous fractures are common in children with spina bifida, especially after a period of immobilisation in plaster. The lower limb bones tend to become decalcified, thin and porotic. When fractures develop they are almost always painless and are only revealed by the development of swelling of the thigh or leg. There is often a mild pyrexia that may give rise to the mistaken diagnosis of osteomyelitis.

Fractures should not be treated by extensive splintage or plaster immobilisation. Union occurs rapidly and the only requirement is light splintage with wool and bandage to maintain the position of the limb.

Renal tract disease

It is not the place of this paper to consider the problems and management of bladder paralysis and renal tract disease that may present in spina bifida. Those concerned with the management of children with spina bifida should, however, always be aware of the possibility of recurrent urinary infection and a liability to develop severe hydronephrosis and renal deterioration that may ultimately lead to death. In the early years of childhood, it is often advisable to express the bladder regularly to prevent bladder distension and reflux. In an older child, and especially in a girl, urinary incontinence can be treated satisfactorily by the performance of ileal conduit operation and this procedure is also valuable when there is evidence of early hydronephrosis to relieve pressure on the renal tract. The conduit may be placed either in the right or left iliac fossa using ileal or sigmoid bowel, to form a suitable conduit. With a satisfactorily applied ileostomy bag, good continence can be obtained and renal function preserved.

Social care and education

Many spina bifida children, if treated adequately from the early days of life, may achieve normal or near-normal lower limb function so that they will be able to attend normal schools and receive normal education. Those with partial paralysis may need to be accommodated in schools for the handicapped but, provided that serious hydrocephalus has been avoided, the intelligence is often good and it is not appropriate to these children that they should be placed in a school such as might be needed for a spastic child with an associated mental handicap. The very severely paralysed child may need to spend a fair proportion of this day in a wheel-chair but it is important that he should continue to receive physiotherapy treatment and encouragement to walk with bracing even over short distances, so as to maintain strength in his limbs and to improve bone texture and cutaneous circulation.

A few severely affected children, probably between 10 and 20% of all those with myelomeningocele, have such a severe spinal lesion associated with such marked hydrocephalus that even the best surgical and medical treatment fails to result in satisfactory mental and physical rehabilitation. It has recently been suggested that there may be room for selection of these children at birth for conservative management in the hope that they may die an early death. Such a decision is always extremely difficult to make and, where there is any doubt, all possible attempts should be made to preserve spinal cord and cerebral function so that the child may at least become able to walk independently.

Summary

The rehabilitation of the child with spina bifida commences on the first day of life. Assessment of the spinal lesion, of lower limb deformity and of the presence of partial or complete lower limb paralysis is made on the first day of life and followed by immediate closure of the spinal lesion. Control of hydrocephalus and urinary complications is undertaken during the first month of life and continued thereafter.

Correction of lower limb deformity should not be made in the neonatal period but delayed until after the third or fourth month of life. Between the third month and the second year of life, operative correction of deformity and tendon transplantation should allow the majority of children with myelomeningocele to become independent in walking, if necessary with the aid of appropriate bracing.

The presence of sensory loss precludes the application of splints or plaster to correct deformity and requires continuous attention to avoid the development of pressure ulceration. Urinary incontinence can be treated by an ileal conduit procedure.

In over 80% of children, satisfactory rehabilitation and education to an independent existence can be assured.

Zusammenfassung

Die Rehabilitation eines Kindes mit Spina bifida beginnt am 1. Tag seines Lebens. An diesem Tag müssen bereits Art und Ausdehnung der spinalen Läsion, der Fehlstellung im Bereich der Beine und Füsse und der neurologischen Ausfälle genauestens erfasst und der Duralsack geschlossen werden. Mit der Behandlung von Hydrozephalus und Harnwegkomplikationen wird im Laufe des 1. Lebensmonats begonnen und später weitergefahren.

Die Korrektur von Fehlstellungen der Beine wird erst nach dem 3. oder 4. Lebensmonat an die Hand genommen. Sie besteht in der Durchführung chirurgisch-orthopädischer Rekonstruktionsmassnahmen einschliesslich Sehnentransplantationen vom 3. Lebensmonat bis zum Ende des 2. Lebens-

jahres. Dadurch wird die Mehrzahl der Kinder mit Myelomeningozele instand gesetzt, ohne fremde Hilfe zu gehen, notfalls unter Anpassung von Stützapparaten.

Liegen Sensibilitätsausfälle vor, so dürfen Gips- und andere Schienen, die sonst zur Korrektur von Fehlstellungen geeignet wären, wegen der Gefahr des Auftretens von Druckgeschwüren nicht angewendet werden. Harninkontinenz kann durch Verpflanzung der Ureteren in den Dünndarm behandelt werden.

Für mehr als 80% dieser Kinder kann eine befriedigende Rehabilitation und Ausbildung zu einer unabhängigen Existenz im späteren Leben gesichert werden.

Résumé

C'est au premier jour de la vie que commence la réhabilitation de l'enfant atteint de spina bifida. C'est en effet au premier jour déjà que l'on peut constater la présence d'une lésion de la moelle, la déformation des membres inférieurs et une paralysie partielle ou complète de ceux-ci, et l'on doit immédiatement refermer la lésion médullaire. Quant à la correction d'une hydrocéphalie ou de complications urinaires, elle se fait au cours du premier mois et sera continuée ensuite.

La correction de déformités des membres inférieurs ne devrait pas se faire dans la période postnatale immédiate, mais devrait être renvoyée au troisième ou quatrième mois après la naissance. C'est entre le troisième mois et le second anniversaire que les corrections opératives des déformités et les transplantations de tendons devraient pouvoir rendre la plupart des enfants atteints de myéloméningocèle assez indépendants pour pouvoir marcher, avec un appareillage approprié si cela est nécessaire.

Des troubles sensoriels excluent l'application de plâtres ou d'éclisses pour corriger une déformité et nécessitent une attention perpétuelle pour éviter un décubitus. Une incontinence urinaire peut être soignée par une anastomose iléale.

Dans plus de 80% de ces enfants l'on arrive à une réhabilitation et éducation satisfaisantes et qui permettent une existence indépendante.

Riassunto

La riabilitazione del bambino che soffre di spina bifida comincia il primo giorno di vita. Infatti già il primo giorno si può constatare la presenza di una lesione spinale, la deformazione dei membri inferiori e una paralisi parziale o completa di questi. È necessario di richiudere immediatamente la lesione midollare. Per quanto riguarda la correzione di un idrocefalo o di complicazioni urinarie, essa deve esser fatta durante il primo mese e sarà continuata in seguito.

La correzione delle deformità dei membri inferiori non dovrebbe essere

fatta durante il periodo post-natale immediato, ma dovrebbe essere rimandata al terzo o quarto mese dopo la nascita. Fra il terzo mese e fino al secondo anno d'età, le correzioni operative delle deformità ed i trapianti dei tendini dovrebbero far sì che la maggior parte dei bambini sofferenti di mielomeningocele diventino abbastanza indipendenti per poter camminare, se necessario con l'aiuto di apparecchi appropriati.

I disturbi che toccano la sensibilità non permettono l'applicazione di gessi o di stecche per correggere una deformità; essi necessitano delle cure continue, al fine di evitare un decubito. Un'incontinenza urinaria può essere curata per mezzo di un'anastomosi ileale.

In più dell'80% di questi bambini si può ottenere una riabilitazione ed educazione soddisfacente, che permette un'esistenza indipendente.

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