Acta orthop. scand. 46, 356–363, 1975

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THE ORTHOPAEDIC MANAGEMENT OF SPINA BIFIDA

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The aims of orthopaedic management are to correct deformity and maintain the correction, to obtain the best possible locomotor function, and to prevent or minimise the effects of sensory loss. Assessment of deformity and paralysis of the lower limbs and trunk needs to be made as soon as possible after birth, at 3-monthly intervals for the first year of life and at 6-monthly intervals until growth is complete. Deformity is often present at birth and may be due to unbalanced muscle action in utero, the effects of posture and intrauterine pressure or congenital maldevelopment, or a combination of any of these. Deformity arising after birth is most commonly due to unbalanced muscle action and sometimes to malunion associated with a spontaneous fracture. Paralysis is due to congenital aplasia or dysplasia of the spinal cord and its roots or to the effects of infection, traction, pressure from lipomata or cysts or vascular damage. The affected muscles may show either normal voluntary activity, voluntary activity with overlying spasticity, autonomous or reflex activity without voluntary control or complete paralysis. A correct assessment of muscle activity that is needed to determine appropriate orthopaedic and surgical management requires clinical assessment of muscle activity combined with electrical stimulation of motor nerves and muscles. Electromyography can be used but is of secondary value.

PRINCIPLES OF ORTHOPAEDIC MANAGEMENT

Conservative methods to correct deformity are inadvisable, especially in the foot because of sensory loss and the risk of producing pressure ulcers. Even if deformity is successfully corrected by conservative means, it is likely to recur if there is persistent muscle imbalance. Deformity can be corrected by elongation of short tendons and muscles and, provided it is done before the age of 2 years, most deformities, even

those that are severe, can be corrected without operating upon bone. Muscle activity needs to be balanced by tendon transfer or by denervation or excision of over-active muscle. If deformity still remains after correction by soft tissue release procedures, further correction by osteotomy or excision of bone and cartilage may be needed, even in young children. Failure to correct deformity, especially in the foot, is likely to lead to serious secondary consequences in later childhood.

Immobilisation or fixation with plaster casts or splints should be kept to a minimum and rarely for longer than 1 month. When deformity has been corrected at all joints, a programme of braces and supports combined with physiotherapy can be used to promote ability to walk with the help of suitable appliances.

The age of the patient is no bar to performance of corrective surgical procedures, but the ideal age at which surgery on the limbs should be commenced is between the sixth month and the second year of life. Between the ages of 2 and 5 years, there may be need for further procedures if deformities recur or alternative deformities develop as a result of modification in muscle activity. After the age of 12, final corrective surgery may be needed, for example to correct deformities of the spine or to arthrodese joints in the foot.

SURGICAL PROCEDURES

Almost every patient presents with a different combination of problems of paralysis and deformity. In many, multiple operations may be needed to correct deformity at the hip, knee, foot and spine such that the total number of orthopaedic procedures needed during childhood averages about eight procedures and in some children may be as many as sixteen. In general, correction of the limbs should be made proximodistally. It is impossible to describe all the procedures that may be needed for the complexity of lesions that may occur, but certain procedures have proved to be specifically indicated in spina bifida.

Hip. Flexion-lateral rotation deformity of the hip is commonly found when there is predominant innervation from the upper lumbar segments with weakness distal to this level. The main active muscles are the ilio-psoas and sartorius muscles and the limb tends to lie in flexion, lateral rotation and abduction. Flexion deformity of more than 30° requires correction by release of short flexor structures and transplantation of the ilio-psoas tendon to the anterior aspect of the greater

trochanter to convert it into a flexor and medial rotator. When this has been done, walking is possible with the aid of extensive bracing.

Flexion-adduction deformity is often accompanied by dislocation of the hip and is associated with innervation from the upper three or four lumbar segments and paralysis distal to this level. It may be present in the child at birth or may develop during the first 2 or 3 years of life.

Correction is ideally accomplished at the age of 9 months and in two stages. At the first stage, a radical division of tight adductor muscles and tendons should restore a normal range of abduction of 80°. Two weeks later, flexion deformity is corrected and the muscle activity balanced by postero-lateral transplantation of the ilio-psoas muscle. At the same time, the hip joint can be opened, the dislocation reduced and capsulorraphy performed. Provided that an adequate reduction has been obtained, subsequent development of the hip is good. Active abduction sufficient to prevent a positive Trendelenburg gait is seldom achieved but, in more than 70 % of patients, walking is possible without bracing at hip level (Sharrard 1964).

If subluxation or dislocation has not been corrected surgically before the age of two, it may be necessary to combine adductor release, open reduction and postero-lateral ilio-psoas transplantation with innominate osteotomy or acetabuloplasty followed, if necessary, by varus intertrochanteric upper femoral osteotomy.

Knee. Deformity at the knee may occur in flexion, extension (recurvatum), valgus, varus or rotation. Flexion or extension deformity is usually the result of muscle imbalance and varus or valgus deformity is more often secondary to a spontaneous fracture.

Flexion deformity is seen most commonly when there is reflex innervation with weakness or complete paralysis of voluntary extension by the quadriceps muscle and spastic activity in the knee flexor muscles. More than 20° of fixed flexion deformity requires surgical correction. In mild cases elongation of the hamstring muscles alone may be sufficient. If deformity is more severe than 20°, it is usually necessary to elongate the short hamstring tendons, to transfer the semitendinosus and biceps tendons to the patella and to correct residual flexion deformity by a supracondylar extension osteotomy with removal of a wedge of bone based anteriorly (Sharrard 1971).

Extension or recurvatum deformity is due to a quadriceps acting strongly in the presence of paralysis of knee flexors with anterior prolapse of the sartorius and gracilis tendons over the medial condyle of the femur. Correction can be obtained by elongation of the quadriceps

by V-Y plasty of the quadriceps tendon (Curtis & Fisher 1969), posterior transfer of the sartorius and gracilis tendon insertions at the tibia and transfer of the adductor magnus tendon to the semitendinosus tendon.

Foot. Almost any possible variety of deformity in equinus or calcaneus of the hindfoot or forefoot, varus or valgus of the hindfoot or forefoot and abduction or adduction of the hindfoot or forefoot may occur. In all but the most lightly paralysed, the soles of the feet are insensitive and are liable to skin breakdown over points of high pressure. There is greater liability to pressure ulceration in the soles of the feet in early adolescence, because increase in the size and weight of the trunk is not matched by growth of the feet. It is essential, therefore, that correction to produce a fully plantigrade foot should be obtained in early childhood to avoid this complication in later childhood. Only a limited amount of correction can be achieved safely by conservative means such as manipulations and plaster casts. Operative treatment can be undertaken at any time between the sixth month and second year of life but, before correction is made, an assessment of activity in the muscles and tendons of the foot must be made, prefarably using electrical stimulation studies.

Equinovarus is the commonest deformity in spina bifida and is usually associated with dominant action of the invertors and plantarflexors combined with weakness of the dorsiflexors and evertors. Even severe deformities can be corrected by elongation of the tendo calcaneus and the tibialis posterior and long toe flexor tendons combined with division of the medial ligaments and capsules of the ankle and subtaloid joints. Muscle balance is restored by lateral transplantation of the tibialis anterior or transplantation of the tibialis posterior through the interosseous membrane to the dorsum and outer side of the foot. Adduction of the forefoot may need to be corrected at a second operation by medial and plantar release of the soft tissues, possibly combined with multiple basal osteotomy of the metatarsals or excision of the cuboid. In severe or recurrent eqiunovarus deformity, especially in an older child, talectomy (Menelaus 1971) may be the only solution; an important step in this operation may be removal of the tip of the fibular malleolus to allow the calcaneus to articulate with the lower end of the tibia.

Paralytic vertical talus deformity is a less common but important deformity of the foot. It is one in which there is equinovalgus of the hindfoot and calcaneo-valgus of the forefoot with dislocation of the talo-navicular joint. The dorsiflexor tendons are short, tight, and

strong, the tibialis posterior is stretched and ineffective, and the intrinsic muscles of the sole of the foot are paralysed. Correction (Duckworth & Smith 1974) needs to be made through two incisions. Through a dorso-medial incision, the tibialis anterior is divided from the navicular, the toe extensor tendons are elongated, the dorsiflexed forefoot is plantarflexed to reduce the dislocation at the talo-navicular joint and reduction is held by means of a transfixing pin. The tibialis anterior tendon is transferred to the neck of the talus. Through a postero-lateral incision, valgus deformity at the hindfoot is corrected by a detachment of the peroneus brevis from its insertion, release of the lateral side of the subtaloid joint, elongation of the tendo calcaneus and transplantation of the peroneus brevis behind the ankle to the medial side to be attached to the navicular at the insertion of the tibialis posterior. If this operation is done before the eighteenth month of life, a good correction can be obtained but, in an older child, it may be necessary to remove the navicular bone in order to obtain correction.

In a limb with complete paralysis below the knee, the foot and ankle tend to fall into valgus. Extra-articular subtaloid arthrosis is inadequate, and pantaloid arthrodesis is contra-indicated in an insensitive limb. Supra-malleolar tibial osteotomy provides a useful means of correcting the valgus deformity (Sharrard & Webb 1974).

The mildest deformity encountered in spina bifida is that of pes cavus and clawing of the toes associated with intrinsic paresis. Deformity of the lesser toes can be corrected without difficulty by transplantation of the long toe flexor to the extensor surface of each toe. Deformity of the great toe can be corrected by a tenodesis of the flexor hallucis longus to the proximal phalanx (Smith & Sharrard 1973) combined with elongation of the extensors of the great toe. The cavus deformity is corrected by release of any tight plantar structures from the calcaneus and from the medial side of the foot.

Spine. Kyphos deformity of the spine is present at birth in one in twenty myelomeningocoeles. If severe kyphosis is present at birth, it may make closure of the spinal lesion difficult or impossible. Although closure can be obtained by spinal osteotomy on the first day of life (Sharrard 1968), this procedure is no longer recommended because of the association with severe paralysis and a liability to early mortality. In some children, kyphosis at birth is mild but increases during growth. Ulceration may develop over the kyphos with difficulty in sitting, increasing paralysis and inability to perform an ileal conduit operation because of compression of the abdominal wall.

In an early case, further progress of the kyphosis may be able to be prevented by a bone graft. Established deformity of more than 90° requires osteotomy-excision of one or two vertebrae of the lumbar spine (Sharrard & Drennan 1972). Correction is seldom perfect, but ulceration can be prevented and upright posture regained. Because of inability to restore the erector spinae to its function as an extensor of the lumbar spine, recurrence of deformity is not uncommon but seldom becomes severe enough to require a second procedure.

Lordosis and lordoscoliosis tend to develop in children with spina bifida at the age of 10 or 11 years, possibly in association with the pre-adolescent growth spurt in which the vertebral bodies grow rapidly without a corresponding growth of the absent posterior bony processes. Corrective measures by means of plaster casts, polythene supports or Milwaukee braces are difficult to apply and likely to cause severe pressure sores in the sacral and iliac regions. Correction can be achieved by skull-femoral traction for 4–6 weeks followed by multiple wedge osteotomy and fusion of the thoraco-lumbar spine using Dwyer staples and cable fixation (Baker & Sharrard 1973).

Scoliosis associated with hemivertebra formation is not uncommonly found in association with spina bifida. If rapidly increasing scoliosis develops in early childhood, vertebral body fusion on the convex side of the curve may be sufficient to prevent further progress of the deformity and correction and further fusion can be made in later childhood or early adolescence.

COMPLICATIONS

The most important complications are those arising as a result of sensory loss. Pressure sores may occur in any area of anaesthetic skin, commonly over areas likely to suffer from pressure or friction over an overlying bony prominence such as the plantar surfaces of the feet or the ischial tuberosities. The child and its parents must be taught to maintain a daily watch on the condition of the skin of the soles of the feet and the ischial areas. Any deformities of the foot must be corrected completely and, if necessary, further correction obtained in adolescence by triple arthrodesis. Footwear specially made to avoid excessive pressure in any area of the sole of the foot may need to be made and, if the child is in a wheelchair, he needs to be taught to avoid sitting on one area of the ischium or perineum for too long at a time.

Spontaneous fractures are liable to occur in the lower limb bones,

especially in those with more extensive paralysis of the lower limbs. The diagnosis is often missed or mis-diagnosed as osteomyelitis. There is swelling of part of the limb with increased heat, a mild general pyrexia, raised sedimentation rate and raised white cell count. Radiographic examination at first may fail to show the fracture, which is often a fracture-separation of an epiphysis, usually at the lower end of the femur. Fractures are particularly likely to occur after a period of immobilisation in plaster. In treatment, plaster fixation or traction with Thomas splintage should be avoided. The simplest measures to maintain the position of the limb are all that is needed and union occurs rapidly with extensive new bone formation.

END-RESULTS

Provided that a child survives beyond the age of 5 years, he has a very good chance of surviving into adult life. The end-result as regards locomotor ability and general function is directly related to the extent of the paralysis and the success of orthopaedic treatment in preventing deformity, to the state of the kidneys and the success of medical and surgical measures in preventing hydronephrosis and renal destruction, and to the presence of hydrocephalus and the success of measures to control it. Provided that correct selection has been made in early life, there can be a reasonable expectation that a child will be able to go into adult life and into suitable and worthwhile employment.

REFERENCES

Baker, R. H. & Sharrard, W. J. W. (1973) Correction of lordoscoliosis in spina bifida by multiple spinal osteotomy and fusion with Dwyer fixation: a preliminary report. *Develop. Med. Child Neurol.* **15**, Suppl. 29, 12–23.

Curtis, B. H. & Fisher, R. L. (1969) Congenital hyperextension with anterior subluxation of the knee. *J. Bone Jt Surg.* **51**-A, 255–269.

Duckworth, T. & Smith, T. W. D. (1974) The treatment of paralytic convex pes valgus. *J. Bone Jt Surg.* **56**-B, 305–313.

Menelaus, M. B. (1971) Talectomy for equinovarus deformity in arthrogryposis and spina bifida. *J. Bone Jt Surg.* **53**-B, 468–473.

Sharrard, W. J. W. (1964) Posterior iliopsoas transplantation in the treatment of paralytic dislocation of the hip. *J. Bone Jt Surg.* **46**-B, 426–444.

Sharrard, W. J. W. (1968) Spinal osteotomy for congenital kyphosis in myelomeningocele. *J. Bone Jt Surg.* **50**-B, 466–471.

Sharrard, W. J. W. (1971) *Paediatric orthopaedics and fractures,* p. 665. Blackwell Scientific Publications, Oxford.

Sharrard, W. J. W. & Drennan, J. C. (1972) Osteotomy-excision of the spine for lumbar kyphosis in older children with myelomeningocele. *J. Bone Jt Surg.* **54**-B, 50–60.

Sharrard, W. J. W. & Webb, J. (1974) Supra-malleolar wedge osteotomy of the tibia in children with myelomeningocele. *J. Bone Jt Surg.* **56**-B, 458–461.

Smith, T. W. D. & Sharrard, W. J. W. (1973) Flexor hallucis longus tenodesis: a new operation for clawing of the hallux. *J. Bone Jt Surg.* **55**-B, 879.

Key words: spina bifida, orthopaedic management of; deformity; sensory loss; osteomyelitis

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