TREATMENT OF SEVERE KYPHOSIS IN MYELOMENINGOCELE BY SEGMENTAL SPINAL INSTRUMENTATION WITH LUQUE RODS

by M. A. C. KADIC and A. J. VERBOUT*

Myelomeningocele leads to kyphosis of the dysplastic spine in 12-20% of cases, resulting in a severe gibbus. In three patients (at the age of 9, 13 and 16 years) with a thoracolumbar kyphosis (90°, 120° and 95° respectively), and a compensatory thoracic lordosis (35°, 105° and 90°) a resection or a wedge osteotomy of the gibbus was performed with segmental sublaminar wire fixation to Luque rods. In addition, a spondylodesis with autogenous bone and an allograft was performed. Correction of the kyphosis (to 30°, 60° and 50°) and lordosis (to 15°, 65° and 55°) was attained. This posterior procedure was sufficient for correction ; there was no need for an anterior release. Cord and dura were left intact. During follow-up (27, 60 and 30 months) no progression of the curves has been noted. This one-stage posterior correction with L-rod fixation proved to be a method of choice for this difficult-to-treat spinal deformity.

Keywords : kyphosis ; myelomeningocele ; Luque rod ; segmental spinal instrumentation.
Mots-clés : myéloméningocèle ; tige de Luque ; instrumentation du rachis.

RÉSUMÉ

M. A. C. KADIC et A. J. VERBOUT. L'instrumentation de Luque dans le traitement des cyphoses graves sur myéloméningocèle.

Dans 12 à 20% des cas, le myéloméningocèle se complique d'une cyphose grave. Celle-ci pourra provoquer une nécrose cutanée, une inclinaison anormale du bassin provoquant une instabilité avec atteinte fonctionnelle des membres supérieurs ainsi que des problèmes thoraciques et abdominaux. Une fixation sublaminaire segmentaire par tige de Luque augmentée de greffes osseuses auto- et homologues fut pratiquée dans 3 cas, âgés respectivement de 9, 13 et 16 ans. En pré-opératoire, il existait une cyphose dorso-lombaire de 95°, 120° et 90° avec une lordose dorsale compensatoire de 90°, 105° et 35°. Deux fois, une résection de la gibbosité fut réalisée et, dans un cas, une ostéotomie cunéiforme. Une désinsertion du ligament longitudinal antérieur ne fut pas nécessaire, une correction suffisante par voie uniquement postérieure étant possible. Une réduction de la cyphose jusqu'à 50°, 60° et 30° fut obtenue. Au niveau dorsal, la lordose fut réduite jusqu'à 55°, 65° et 15°. En post-opératoire et au moment du follow-up (27, 60 et 30 mois), nous ne constatons ni complication importante ni augmentation des courbures. Il n'y eut pas de problème neurochirurgical grâce à l'intégrité de la moelle et de la dure-mère. Nous estimons que la correction postérieure en un temps avec fixation par tige de Luque est un traitement de choix pour cette pathologie grave.

* Orthopedic Department, Leiden University Hospital, Leiden (The Netherlands).

SAMENVATTING

M. A. C. KADIC en A. J. VERBOUT. Behandeling met het Luque instrumentarium van ernstige kyfose bij myelomeningocele.

Myelomeningocele leidt in 12-20% tot een kyphose van de dysplastische wervelkolom. Deze kan leiden tot een ernstige gibbus met huidproblemen, zitinstabiliteit met daardoor functieverlies van de armen en tenslotte problematiek inzake de organen in borst en buikholte. Segmentale sublaminaire draadfixatie aan Luque staven met autologe en homologe botttransplantatie werd toegepast bij drie patiënten van 16, 13 en 9 jaar, met een thoraco-lumbale kyphose van respectievelijk 95°, 120° en 90°. Craniaal hiervan was een compensatoire thoracale lordose van 90°, 105° en 35° aanwezig. Tweemaal werd een resectie van een deel van de gibbus en eenmaal een wigosteotomie verricht. Het ventraal vrijmaken was niet noodzakelijk ; voldoende verschuiving en adaptatie was mogelijk tijdens de dorsale benadering. Een correctie tot 50°, 60° en 30° werd bereikt, terwijl thoracaal de lordose tot 55°, 65° en 15° werd geredresseerd. Belangrijke postoperatieve complicaties traden niet op, evenmin als progressie van de curven in de follow-up (27, 60 en 30 maanden). Neurochirurgische problemen zoals intradurale druktoename of liquor lekkage cq. fisteling werden niet gezien hetgeen samenhangt met het intact houden van ruggemerg en dura. Deze eenmalige posterieure correctie en fixatie met Luque staven verdient de voorkeur in de behandeling van de ernstige kyphotische wervelkolom deformiteit bij meningomyelocele.

INTRODUCTION

The incidence of spina bifida cystica differs among countries ; in the Netherlands it occurs in 1.2 of 1000 births. In about 90% it involves a myelomeningocele, which means that neurological disturbances due to the myelodysplasia are common. The life expectancy of children born with severe myelomeningocele has clearly improved over the last 2 decades, due to advances in primary neurosurgical treatment of the meningocele, in prevention and/or treatment of infection, and in the care of the secondary problems associated with hydrocephalus. However, during growth many problems may have time to arise (13). From the orthopedic point of view, in cases with higher neurosegmental deficits, the stability of the spine and the balance of the trunk have become increasingly significant and difficult problems.

It is essential to treat the spinal deformities adequately to maintain the pelvic stability or to preserve the benefits of previous treatment of lower limb disturbances (such as contractures, fractures or dislocation of the hip). Because many of these children suffer from cerebellar dysfunction leading to coordination problems in the upper limbs, they need both hands to perform simple tasks, and cannot afford the loss of the use of one upper extremity ; having to use one hand to maintain equilibrium because of imperfect sitting balance leads to an extra handicap. Spinal deformities may occur in the sagittal or frontal plane, so a kyphotic, lordotic, or scoliotic curve may appear. In the sagittal plane these problems may be particularly difficult and require special attention.

CLASSIFICATION

Kyphotic and/or lordotic deformities may be classified as congenital or paralytic (21). The congenital type is caused by anomalous vertebral development in addition to the spina bifida. The deformity is already present at birth. In 10% of cases an obvious angular gibbus, rigid from the start, is present at birth (9) (fig. 1). It is usually located in the upper lumbar spine (12). In the first years of life the kyhotic curve progresses markedly. The structures on the anterior side are thought to shorten significantly. Furthermore, a tendency towards a compensatory lordosis, above the level of the kyphosis, quickly appears.

The paralytic type does not have an obvious deformity at birth, except the apert spina. Owing to muscle imbalance, habitual posture and the inclination of the pelvis, a kyphotic deformity develops in nearly a third of patients by early adolescence (20). The gibbus is more arched with less rigidity and primary shortening of the anterior structures than in the congenital types.

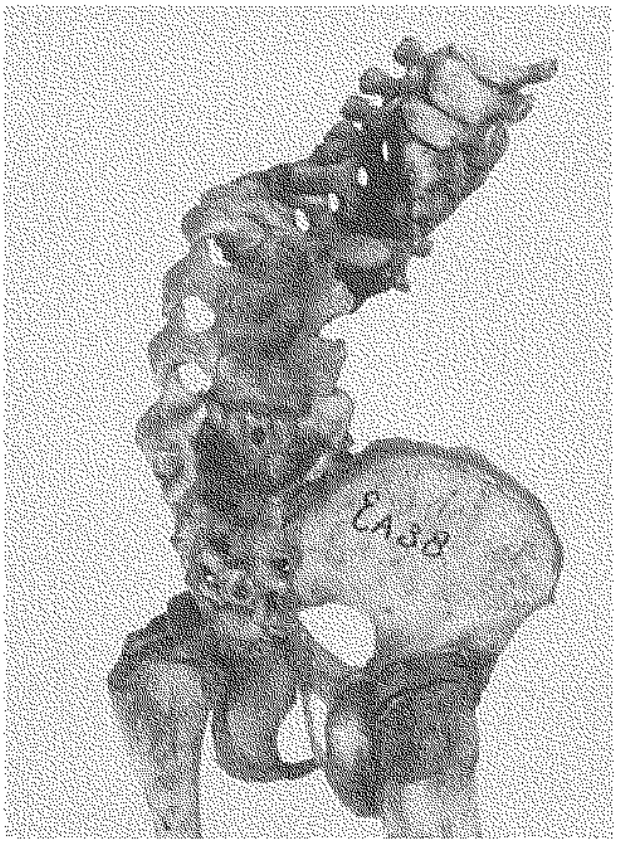


Fig. 1. — Axial skeleton of fetal specimen with spina bifida. In the upper part the arches are closed ; more caudally, in the region of the defect, a severe kyphosis is present. The transverse processes point laterally, in a normal fashion ; nevertheless the foramina are visible between the pedicular structures.

PATHOGENESIS

In newborn children with spina bifida cystica and congenital kyphosis, instead of being posterior to the spine, the erector spinae and quadratus lumborum muscles are laterally displaced and thus cause flexion mechanically (4, 10). Furthermore, the transverse processes are thought to be pulled ventrally by the psoas muscles, leading to ventrolateral migration of the thoracolumbar fascia, which enhances the progression of the kyphosis (see fig. 2).

Cranial to the kyphosis, where a more adequate musculature is generally present, a compensatory thoracic hyperextension develops, leading to thoracic hyperlordosis.

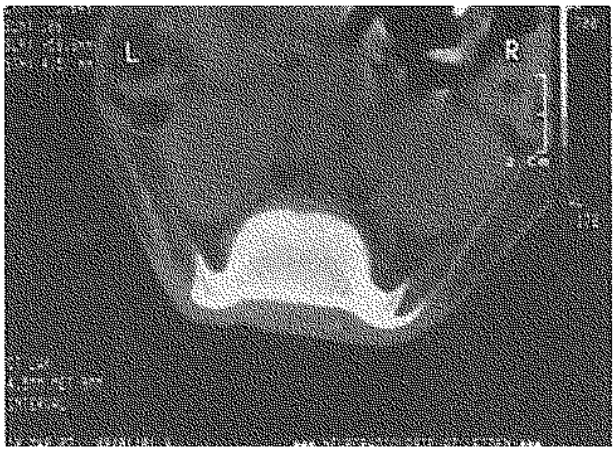


Fig. 2. — Case 3. CT scan at the level of the apical vertebra of the gibbus. The lateral insertion as well as orientation of pedicles, transverse processes and arch rudiments, are shown.

TREATMENT

The application of braces is difficult, at least in part because of the risk of pressure sores over the anesthetic skin. Furthermore, despite braces, the curves commonly progress, due to the progressive nature of the process and the strong deformative forces. Operative therapy is therefore inevitable. Delay of surgery is preferable to resection and fusion at a too young age, because the fragile bone lacks sufficient strength to withstand instrumentation. In addition, long fusions are undesirable because of the resultant shortening of the torso which may lead to depression of respiratory function.

Since Sharrard advocated surgical treatment in 1968, the procedure has evolved because of progress in the development of instrumentation. Previous techniques with inadequate instrumentation were associated with several complications such as material breakage, pseudarthrosis, recurrent kyphosis and extreme osteoporosis with lower limb fractures due to prolonged immobilization (1, 3, 5, 6, 9, 12, 14, 16, 17, 18, 19, 22).

Indications for surgical treatment

There are several indicators for surgical treatment of kyphosis in myelomeningocele. Chronic re-

current ulceration of the skin, which can lead to episodes of meningitis, is a frequent indication. Fixed flexion of the pelvis and prominent kyphosis may make the fitting of walking aids impossible. The increased flexion of the runk also may interfere with a surgical procedure such as ureterostomy, ileostomy or vesicotomy. Urinary diversion may be necessary because of hydronephrosis, ureteral reflux or chronic urinary infections. On the other hand, care of an already existent stoma may become impossible. Progressive deterioration of motor and sensory activity may occur cranially to the paraplegic level because of the increasing extension of the spinal cord over the kyphosis or because of the presence of diastematomyelia and a tethered cord. The grotesque deformity and abnormal posture may in themselves be an indication. Other less frequent indications are pain or increasing respiratory restriction.

CASE REPORTS

1. A boy was born in 1971 with total paralysis below the level of L1. Two days after birth a thoracolumbar myelomeningocele was neurosurgically treated.

A paralytic kyphosis of 60° at the level of L1-L3 was noted in 1972. Over the years, a rapid progression of the curve and a compensatory thoracic lordosis developed (fig. 3). In 1984 the kyphosis between T12 and L4 and the compensatory lordosis between T1 and T8 measured 120° and 105°, respectively. The sitting balance had been disturbed with loss of function of the hands. Manual bladder expression became difficult, and skin problems over the kyphosis occurred. Two weeks preoperatively, at the age of 13.5 years, a halotraction was applied in order to make the spine more supple (see fig. 6A) ; correction was achieved especially in the lordotic part. At operation a posterior resection of the upper part of the gibbus was performed, with fixation to Luque rods using segmental wiring from level T3 to the sacrum (fig. 4). The rod fixation was extended through the pelvic cristae (transiliac). Autologous bone from the resected gibbus and allografts completed the posterior spondylodesis. The dura was partially detached but not excised. Correction measured 60° and 65°, respectively.

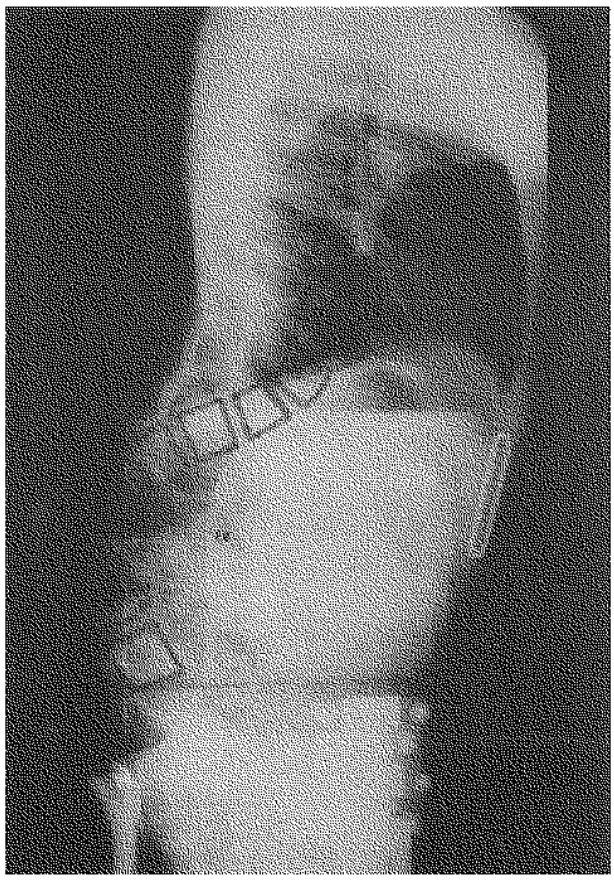


Fig. 3. — Case 1. Kyphosis with wedge-shaped flattening of the apical vertebrae accompanied by a compensatory thoracic lordosis.

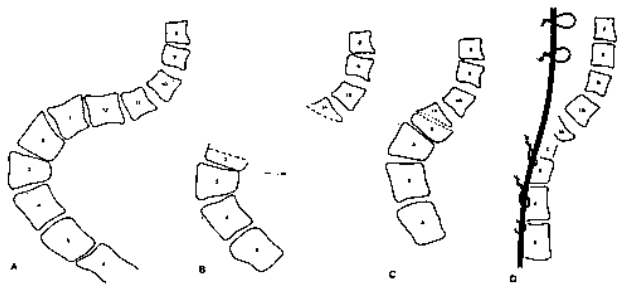
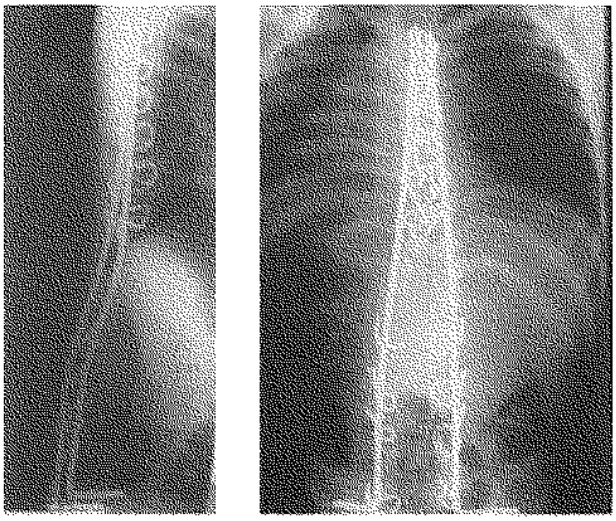


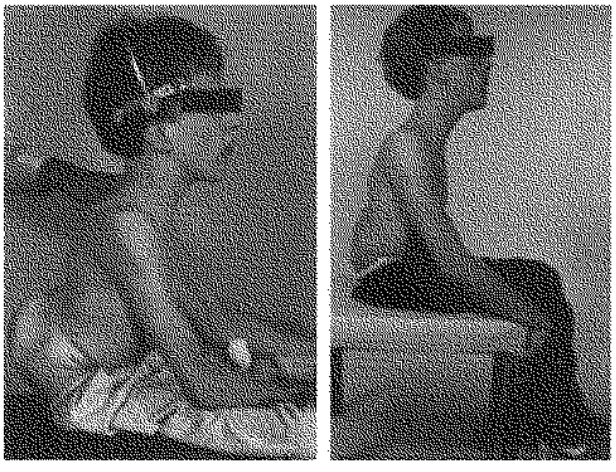
Fig. 4. — Case 1. Schematic representation of the technique of gibbus resection (6) followed by fixation.

An individually molded total body contact TLSO* composed of 2 parts connected by a zipper was applied for 6 months. With the patient in a horizontal position, one part can be removed, simplifying hygienic care. After a 60-month follow-up, the objective and subjective results are good. The internal fixation is intact without complications on X-ray ; no signs of progression of the curve have occurred (figs. 5 and 6).



a b

Fig. 5. — Case 1. Postoperative lateral (A) and AP (B) roentgenographs.



a b

Fig. 6. — Case 1. Preoperative clinical photograph (A) and postoperative situation (B) after 54 months' follow-up.

2. A girl born in 1970 was paralyzed below the T12 level. Her case was similar to patient 1. The kyphosis (T12-L3) and lordosis (T3-T12) measured 95° and 90°. The indication for gibbus resection was sitting imbalance, worsened by the right and left convex scoliotic curve of 95° (T2-T12) and 35° (T12-L4), resulting in gluteal pressure sores. Respiratory capacity was restricted and recurrent bronchitis occurred. Technically the operation was identical to that for patient 1, as was the postoperative treatment. The correction of kyphosis and lordosis was 50° and 55°, and of scoliosis, 35° and 10°. No complications (30 months follow-up) have arisen so far.

3. A girl born in 1978 with paralysis below the level of T12 had a thoracolumbar myelomeningocele that epithelialized spontaneously. An initial kyphosis of 15° at the level T11-L5 progressed and led to severe skin problems. To avoid wound problems after the planned spinal correction, plastic surgery was carried out 6 months in advance, using transposition of the latissimus dorsi and gluteus maximus muscles, followed by a split skin graft 2 months later. In 1987 the kyphosis (T11-L5) measured 90°, whereas the lordosis (T3-T11) was 35°. The sitting balance was seriously disturbed. A posterior wedge columnotomy was performed at level L1-2 ; Luque-rod fixation and sublaminar wiring were used. Kyphosis and lordosis were corrected to 30° and 15°. After a 27-month follow-up the sitting balance is perfect ; no skin ulcerations and no further progression of the curve have occurred.

DISCUSSION

Twenty years, several techniques have been used to attempt fusion and fixation of a kyphotic spine in paraplegic patients with a myelomeningocele (1, 3, 5, 6, 7, 8, 9, 11, 12, 14, 15, 17, 18, 19, 21, 23). However, the failure and recurrence rate have been high, caused by the insufficient fixation in the relatively osteoporotic bone, in the absence of stabilizing posterior structures of the spine. Seg-

* TLSO = Thoraco-Lumbal-Sacral Orthosis ; Neofract® trunk orthosis.

mental spinal instrumentation according to Luque, with fixation on several levels, is a new and apparently good technique in this type of pathology (1, 2, 9, 15, 23). In the spina bifida area the wiring is passed through the intervertebral foramina or through predrilled holes in the vertebral body or pedicles.

In our 3 patients, the extensive anterior release and/or spondylodesis, recommended by other authors (1, 3, 11, 16), was not necessary. In patients 1 and 2, suffering from the most serious kyphosis and compensatory lordosis, the gibbus was indeed reduced by approximating the two parts of the spine in an antero-posterior direction rather than by correcting the gibbus itself (see fig. 4). This also corrected the lordosis. In parient 3, where the compensatory lordosis was not as marked, sufficient correction was obtained using a wedge osteotomy at the level of the apical vertebra without section of the anterior longitudinal ligament. In our experience, a satisfactory correction can be obtained by only the posterior procedure. Contrary to some authors (7, 9, 21, 23), we believe that a nonfunctional cord should not be resected because this may modify the hydrodynamics of the cerebrospinal fluid and lead to increased intracerebral pressure if drainage is not optimal (24). Persistent leakage of fluid may occur after resection with an increased risk of meningitis.

CONCLUSION

Use of the posterior approach alone is sufficient for correction ; a prior extensive anterior release seems superfluous. A compensatory thoracic lordosis could, and should, be corrected during the same stage. Instrumentation with the Luque segmental rod system, in combination with bone repair, has resulted in stable fixation without later progression of the kyphosis. There is no need for a resection of any neural structures ; maintaining an intact myelum decreases the complication rate.

Acknowledgments

The senior author (A. J. Verbout) wishes to thank his fellow members of the Spina Bifida Team (University of Leiden), especially Dr. J. H. C. Voormolen, neurosurgeon, and Prof. B. D. de Jong, plastic surgeon. He is greatly indebted to Dr. W. Keessen, orthopedic surgeon, Utrecht, who assisted with the operations.

REFERENCES

1. BANTA J. V. The evolution of surgical treatment of spinal deformity in myelomeningocele. *Z. Kinderchir.*, 1987, *42*, Suppl. I, 10-12.
2. BROOM M. J., BANTA J. V., RENSHAW T. S. Spinal fusion augmented by Luque-rod segmental instrumentation for neuromuscular scoliosis. *J. Bone Joint Surg.*, 1989, *71-A*, 32-44.
3. BROWN H. P. Management of spinal deformity in myelomeningocele. *Orthop. Clin. North Am.*, 1978, *9*, 391-402.
4. DRENMANN J. C. The role of muscles in the development of human lumbar kyphosis. *Dev. Med. Child Neurol.*, 1970, **12**, suppl. 22, 33-38.
5. ECKSTEIN H. B., VORA R. M. Spinal osteotomy for severe kyphosis in children with myelomeningocele. *J. Bone Joint Surg.*, 1972, *54-B*, 328-333.
6. EYRING E. J., WANKEN J. J., SAYERS M. P. Spine osteotomy for kyphosis in myelomeningocele. *Clin. Orthop.*, 1972, *88*, 24-31.
7. HALL J. E., POITRAS B. The management of kyphosis in patients with myelomeningocele. *Clin. Orthop.*, 1977, *128*, 33-40.
8. HELLINGER J. Die Resektionskolumnotomie bei der myelodysplastischen Lumbalkyphose, *Pädiatr. Grenzgeb.*, 1981, *20*, 313-322.
9. HEYDEMANN J. S., GILLESPIE R. Management of myelomeningocele kyphosis in the older child by kyphectomy and segmental spinal instrumentation. *Spine*, 1987, *12*, 37-41.
10. HOPPENFELD S. Congenital kyphosis in myelomeningocele. *J. Bone Joint Surg.*, 1967, *49-B*, 276-280.
11. LEATHERMAN K. D., DICKSON R. A. Congenital kyphosis in myelomeningocele. *Spine*, 1978, *3*, 222-226.
12. LINDSETH R. E., STELZER L. Vertebral excision for kyphosis in children with myelomeningocele. *J. Bone Joint Surg.*, 1979, *61-A*, 699-704.
13. LORBER J. Results of treatment of myelomeningocele. *Dev. Med. Child Neurol.*, 1971, *13*, 297-303.
14. LOWE G. P., MENELAUS M. B. The surgical management of kyphosis in older children with myelomeningocele. *J. Bone Joint Surg.*, 1978, *60-B*, 40-45.
15. LUQUE E. R. The correction of postural curves of the spine. *Spine*, 1982, *7*, 270-275.
16. MAYFIELD J. K. Severe spine deformity in myelodysplasia and sacral agenesis. An aggressive surgical approach. *Spine*, 1981, *6*, 498-509.
17. McMASTER M. J. The long-term results of kyphectomy and spinal stabilization in children with myelomeningocele. *Spine*, 1988, *13*, 417-424.

18. SHARRARD W. J. W. Spinal osteotomy for congenital kyphosis in myelomeningocele. *J. Bone Joint Surg.*, 1968, *54-B*, 466-471.
19. SHARRARD W. J. W., DRENMANN J. C. Osteotomy-excision of the spine for lumbar kyphosis in older children with myelomeningocele. *J. Bone Joint Surg.*, 1972, *54-B*, 50-60.
20. SHURTLEFF D. B., GOINEY R., GORDOW L. H. Myelodysplasia : The natural history of kyphosis and scoliosis : a preliminary report. *Dev. Med. Child Neurol.*, 1976, *18*, supp. 37, 126.
21. SLOT G. H. *De spondylodese volgens Dwyer bij ernstige wervelkolom deformaties.* Proefschrift Nijmegen (1980), p. 36-39, p. 75, 135.
22. SRIRAM K., BOBECHKO W. P., HALL J. E. Surgical management of spinal deformities in spina bifida. *J. Bone Joint Surg.*, 1972, *54-B*, 666-676.
23. STEPHEN J. P., BODEL J. G. Luque rod fixation in meningomyelocele kyphosis : a preliminary report. *Aust. NZ. J. Surg.*, 1983, *53*, 473-477.
24. WINSTON K., HALL J., JOHNSON D., MICHELI L. Acute elevation of intracranial pressure following transection of non-functional spinal cord. *Clin. Orthop.*, 1977, *128*, 41-44.

M. A. C. KADIC
Department of Orthopedic Surgery
A.Z.L. (University of Leiden)
Postbus 9600, 2300 RC Leiden
(The Netherlands)