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Sagittal static imbalance in myelomeningocele patients: improvement in sitting ability by partial and total gibbus resection

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Abstract The progression of kyphosis in myelomeningocele is independent of skeletal growth and requires early operative correction and stabilization to prevent a loss of sitting ability. In severe cases, only vertebratomy makes it possible to achieve correction, stability and skin-closure without tension. In 14 patients with myelomeningocele gibbus, kyphectomy was performed, removing two vertebral bodies on average. The average kyphosis angle decreased from 128° to 81°, enabling most of the patients to participate again in social life by restoring wheelchair mobility. Nevertheless, a significantly higher complication rate was found compared to other correc-

tional operations, lengthening the average hospital stay to 41 days. Special problems arose from trophic disorders of the skin and soft tissue and from the dystrophic muscles below the level of neural malfunction. In three cases, kyphosis reappeared cranial to the fused segments, requiring ventral stabilization. With respect to increasing kyphosis angle, an early intervention should be aimed at. A secondary operation can be necessary, if surgery is performed without taking care of the growth potential.

Key words Myelomeningocele · Sagittal imbalance · Kyphosis · Sitting ability

Introduction

The myelomeningocele (MMC) is one of the dysraphic disorders of the neural arch. Pathogenically it is caused by the absence of proliferation of neuroectodermal cells after migration is finished. The occult form without neurologic deficit is estimated to have incidence of up to 1%, while a spina bifida aperta cystica with everted myelon and complete paraplegia occurs in 0.005% of live births. With the help of postnatal closure of the sac and the early commencement of hydrocephalus therapy, the life expectancy of patients with myelomeningocele has nearly reached the level of the normal population [3, 24]. While therapy during infancy concentrates on taking care of the cerebral and renal function, during growth and adolescence, more and more problems concerning the skeletal system arise, caused by the wish for maximum mobility and improved nursing

aspects such as lying in supine position and sitting without support. The clinical appearance is dominated by the spastic or paralytic neurological lesion at different levels. In addition, the increasing spinal deformity in combination with trophic skin and soft tissue disturbances leads to secondary problems, i.e. decubital ulcers and contractures of hips and limbs. Kyphosis appears in 12–28% of the cases, with neurologic damage at the thoracic level [2, 15, 24, 25]. The gibbus, however, develops usually between L2 and L5 [4].

Problems of the kyphotic spine

Myelomeningocele at the thoracic level is in most cases accompanied by other malformations of the neural arch. They can aggravate each other, if for example during progressive kyphosis the dura is pulled caudally by a tethered

cord. Brainstem and cerebellum can be displaced into the foramen magnum, as in Arnold Chiari Malformation, with disturbance of cerebro-spinal fluid (CSF) circulation and secondary neurological complications. Skin alterations over the prominent apex of the gibbus caused by braces or wheelchair lead to ulcers and fistulae, which can only be insufficiently controlled because of the trophic disorders. Continuing progression of kyphosis finally ends up in a complete loss of upright posture and sitting ability. As a consequence, patients who are already wheelchair bound get totally deprived of their mobility. Such patients, despite normal intelligence, are likely to suffer restrictions on their social integration.

Therapy

Conservative orthopedic measures, like night time braces and walking apparatus, cannot prevent structural deformities. Especially in wheelchair-bound patients, the chance of developing a scoliosis or kyphosis is high, the probability increasing with the level of motor malfunction. It has to be kept in mind, that these deformities remain progressive after stoppage of growth, in contrast to idiopathic scoliosis [3, 9].

Indication for kyphectomy

There is no recommendation regarding a critical angle in the sagittal plane in cases of kyphotic spine. Simultaneous kyphectomy with postnatal closure of the sac is proposed by Lindseth 1994 [17], but seldom performed [3]. Rapid progression during the growth spurt forces the patients into orthopedic care. The patient with the least extensive kyphosis referred to the orthopedic department showed a sagittal angle that had already reached 80°. Additional indications for surgical intervention beside spinal deformity therefore are:

1. Reduced sitting ability
2. Persisting sores at the apex

Material and methods

Patients

Between 1993 and 1996, 14 patients suffering from MMC with gibbus were treated by surgical correction. The ratio of girls to boys was 5:4. The average age at the time of first kyphosis correction was 11 years 6 months (4–20 years) (Table 1). The average kyphosis angle was 128° (80°–175°), with nine patients having a scoliotic component greater than 10°. The average scoliosis angle was 36° (0°–97°). The levels of motor deficit were between T5 and L1, with a radiological localization of the first open arch between T7 and L4. A tethered cord was diagnosed by MRI in 13 patients. All of the patients had undergone postnatal ventriculoperitoneal (VP) bypass; only one did not have to undergo surgical closure of the

Table 1 Correction: technique and results in 14 patients (CD Cotrel-Dubousset instrumentation implanted from lateral)

Pat. no.	Age (years)	Sagittal angle	Removed vertebra	Corrected angle	Method
1	4	115°	L2	85°	Wire, screw
2	5	165°	L3, L4	115°	Wire, screws
3	8	160°	L4, L5	90°	Screws
4	9	100°	L1, L2, part L3	28°	Wire
5	10	167°	L3–L5	70°	Screws
6	11	120°	L2	100°	CD
7	12	160°	L2–L4	115°	Screws
8	12	80°	L2	63°	CD
9	12	80°	L3	73°	CD
10	13	140°	L4	90°	Wire, screws
11	15	175°	L1–L4	75°	Screws
12	15	97°	T9–L1	53°	CD
13	15	85°	L2	70°	CD
14	20	150°	L5, part L4, S1	115°	Screws
Mean	11.5	128.1°		81.6°	

myelomeningocele, because of spontaneous occlusion. Further surgery concerned revision or change of the VP shunt, two times on average. Three patients suffered from decubital ulcers before correction. Wheelchair-bound patients complained of a sitting disability because of the gibbus. Most of them had changes to the wheelchair's back and seat, the sitting position being displaced to the front of the seat.

Technique and instrumentation

Kyphosis correction was achieved by resection of two vertebral bodies on average (Table 1). In 13 patients with a tethered cord in addition, transverse myelotomy was performed to prevent an acute insufficiency of liquor circulation (Fig. 1).

Fusion area

In order to leave as much growth potential as possible, resection enclosed the apex and the horizontal vertebral bodies. Uni- or bisegmental spondylodesis was assumed to be stable enough because of simultaneous correction of the sagittal profile. Only in cases with removal of L5 was the pelvis fused. Compensatory thoracic lordosis was neither resected nor fused.

Instrumentation

Of the patients younger than 10 years with kyphosis angles over 100° (100°–175°) (Figs. 2, 3), the resulting angle allowed stabilization using transcorporeal screws ($n = 5$) (Figs. 4, 5). In one case, an intertransverse tension wire band was implanted. In three cases, the two procedures were combined. In older patients with sufficient vertebral stability and angles below 100° (Fig. 6), extensive correction left no segmental angulation and Cotrel-Dubousset (CD) instrumentation was used, being implanted from lateral because of the lack of covering soft tissue (Fig. 7). One patient with a kyphosis angle of 120° was corrected to 100° by lateral CD fixation. Bone grafting was done using spongiosa from the resected parts.

Fig. 1 a–h Schematic illustration of gibbus correction. **a** Myelomeningocele (MMC) gibbus with apex at T12 and paralysis level at T11. After preparation, transversal myelotomy is performed (**b**), and the vertebrae of the gibbus are resected to achieve a tension-free skin closure (**c**). The vertebrae left are then put onto one another (**d**). The choice of instrumentation depends on kyphosis angle, soft tissue situation and grade of deformity. It can be achieved by transcorporeal screws (**e**), dorsal CD instrumentation (**f**), tension wire band (**g**) or lateral CD instrumentation (**h**)

Results

Correction

Performing kyphectomy, the kyphosis angle was reduced by 47° to 81° (range 10° – 115°) on average. Divided by method, the group with intercorporeal screws had an average correction of 69° , from 162° to 93° , resecting 2.6 vertebrae. The patient with a wire loop was corrected from 100° to 28° , removing 2.5 vertebrae. Using wire loop and screws in combination, the kyphosis angle decreased from 138° to 97° ; 1.3 vertebral bodies being resected. Using CD instrumentation, there was a correction from 89° to 67° , with 1.6 vertebrae removed. The scoliosis decreased from 36° to 23° .

Hospitalization data

Intraoperative and postoperative blood loss amounted to 1194 ccm (range 500–2050 ccm) on average. The duration of operation was about 216 min (range 75–240 min). The average stay in hospital was prolonged by the amount of healing disturbances and lasted about 41 days (range 20–81 days). All of the patients were provided with a polyethylene brace as soon as the wounds allowed. Once the spine was corrected, an adaptation of the wheelchair to a more normal profile was possible.

Complications

In six patients primary, and in a further four patients secondary, ulcers over the apex of the gibbus appeared. Four patients had to undergo MESH-graft or flap-shift operation. The secondary sores were treated by partial or total removal of the instrumentation. In four patients, a change of instrumentation was necessary. In these cases of revision surgery, the Luque technique was used in one, and an external fixateur in another. In two cases, lateral CD instrumentation was used, with good results. Loss of correction was noted only in one case. In three cases, the original

Fig. 2 A 14-year-old boy with severe gibbus accompanied by skin alterations after (*left*) and before (*right*) spinal surgery

Fig. 3 Radiograph of the same patient as in Fig. 2, showing a kyphosis with gibbus, with a sagittal angle of 170°

Fig. 4 Same patient as in Figs. 2 and 3. Radiograph shows correction to 75° by vertebrectomy and transcorporeal screw fixation

Fig. 5 Same boy at the age of 15.6 years after removal of the screws. A stable bony spondylodesis is evident without loss of correction

Fig. 6 A 12-year-old patient with a light scoliotic component of 22°

Fig. 7 Same patient as in Fig. 6. Radiographs shows correction by bilateral CD instrumentation; the frontal scoliosis angle is unchanged

correction was maintained or even improved. As another consequence of skin breakdown, four patients showed fistulae above the metal, followed by loosening of the osteosynthesis material. There were no neurological complications (Tables 2, 3).

Table 2 Postoperative complications

Delayed wound healing	6 cases
Septic or aseptic instrumentation failure/loosening	5 cases
Subsequent kyphosis cranial to spondylodesis	3 cases

Table 3 Re-interventions

Partial or complete removal of instrumentation	3 cases
Secondary plastic surgery	3 cases
Change of instrumentation	2 cases
Second intervention with lengthening of fusion	2 cases

Follow-up

The average follow-up was 37.7 months (range 12–75 months). The patients were examined clinically and radiologically at intervals of 1 year. During this time, three cases of progressive secondary kyphosis because of deterioration cranial to the fused segments appeared. These patients had been primarily operated at an age below 8 years. All of them were stabilized by ventral fusion with Cotrel-

Fig. 8 A very young patient subjected to correction by tension wire band at the age of 5 1/2 years. Radiographs show hyperlordosis of the segments cranial to the fusion

Fig. 9 Same patient as in Fig. 8. At the age of 9.9 years, growth-related kyphosis cranial to the fused area is apparent (*left*). At the age of 10, the patient underwent secondary ventral stabilisation by CDH instrumentation and an autologous fibula graft (*right*)

Dubousset-Hopf (CDH) instrumentation, one in combination with a fibula graft (Figs. 8, 9). In two of these patients, the originally achieved correction could be restored, the third suffered a loss of correction from 28° to 50° . In five of seven patients who had to undergo change of instrumentation, the originally achieved correction was re-established. An improvement of 13° and 43° respectively was obtained in two patients.

Altogether, the average corrected kyphosis angle was 79° , only one patient lost his sitting ability during the observation period.

Discussion

Myelomeningocele is classified according to the level of paralysis [23]. In the literature, follow-up studies [3] show good prediction of spinal deformity according to the level of motor malfunction. If the paralysis is at the thoracic level, 83–100% of the patients develop scoliosis or kyphosis [9, 19, 23, 25]. In cases where the neurological lesion is at the sacral level, only about 5% of patients will suffer spinal deformity [23]. The lumbar myelomeningocele generally leads to scoliosis, small side differences in the motor malfunction generating a noticeable sideways displacement of the trunk. Kyphosis is to be seen almost exclusively in myelomeningocele at the thoracic level. This is because of the longer lever of the ventral muscles, which are innervated from more cranial segments [5], and the lateral position of the erector spinae muscles as well as the psoas and quadratus lumborum muscle, with portions even

anterior to the axis of flexion, which produce a marked kyphosis [6, 15].

Conservative brace treatment [24, 26] has been superseded by surgical correction and stabilization, because the sagittal deformity tends to progress, depending on age and paralysis level, despite optimal prevention treatment by brace or other therapies [3]. Follow-up studies by Martin et al. [20] demonstrated prevention of kyphosis progress and skin breakdown by kyphectomy in contrast to non-operative treatment.

The recommendation of the study group for spinal deformities in the DGOT (German Orthopedic and Traumatologic Association) [14] for treatment of MMC is early intervention with correction and fusion. Because of spinal growth, which would be expected to lead to cranial instability, the preferred time for surgery should be after the age of 10. In severe cases, an earlier intervention may be necessary to prevent severe deterioration. From 1968 on, a variety of surgical procedures has been described [1, 5, 7, 8, 10, 21, 24, 26–29]. Lindseth [17] recommends a simultaneous gibbus resection without instrumentation together with postpartal closure of the sac. In older children, instrumentation is necessary to maintain correction and to provide fusion of the adjacent segments.

While scoliosis surgery is performed according to the principles of correction of sagittal and rotational deformity, either from ventral or dorsal approaches, the correction of severe kyphosis with gibbus can require the resection of one or more vertebrae. Because this always takes place below the level of motor malfunction, neurological status is mostly not influenced. Various procedures for correction were used, depending on the age and growth potential of the patient, on the deformation angle in the sagittal and in the frontal plane, and on the size and stability of the vertebral bodies.

In cases of dorsal instrumentation, techniques were preferred in which the instrumentation did not project dorsally above the spinal profile. Extensive correction was achieved with good functional results. When scoliosis correction was

performed at an early age, meaning a less severe deformity, the risk of complications was noticeably lower. Nevertheless, in three patients (21%), the spine cranial to the fusion turned out to be unstable and required extension of the fused area. This happened especially in patients who had undergone correction very early (5–8 years), so a large growth potential was left. This has also been confirmed in long-term follow-up studies by Lintner and Lindseth [18], who saw loss of correction in 34 of 39 patients operated at the average age of 6.10 years. In such cases, correction of the thoracic hyperlordosis with a longer fusion and anterior approach should be considered. Heydemann and Gillespie [10] had only one loss of extensive correction in 12 older children, performing anterior fixation to the pelvis. Banta [1] also reports a correction of 79° and 39° respectively by combined anterior and posterior fusion. Among our patients, anterior fusion was added in the three cases of cranial instability described above, and this correction was maintained over the observation period.

In earlier studies, good primary kyphosis correction of 51° (45%) was achieved, but pseudarthrosis occurred in 67% and infection in 45% of the cases [15]. Combined procedures are also shown to have a pseudarthrosis risk, of 5.7% [1]. Performing kyphectomy, kyphosis angles of up to 170° could be treated with similar good results (up to 100° correction) (Figs. 3, 4). Among the patients described above, no pseudarthrosis was seen. Early loosening in five patients (38%) was caused exclusively by wound healing problems, which were distributed equally among the different fusion methods.

The complication rate was significantly increased compared to other correctional operations. This was mainly due to the trophic skin and soft tissue disturbance, giving rise to delayed wound healing in 64%. It was not correlated to operation time. The duration of operation in MMC scoliosis surgery takes 65% (315 min) more than in correctional

surgery of idiopathic scoliosis [9], the blood loss being more than double (2520 ccm). Among the kyphosis patients, the duration was intermediate, at 216 min, with a blood loss of 1194 cc.

Conclusions

Regarding the almost normal life expectancy of myelomeningocele patients, it is important to maintain sitting ability and wheelchair mobility that allows these patients to participate in social life.

Surgical correction and stabilization is inevitable in established MMC kyphosis, because of the inevitable progression of deformity.

Accompanying problems like hydrocephalus, tethered cord and pressure sores above the gibbus have to be considered when choosing the surgical procedure.

Resection of the gibbus reduces the ventral overhang and improves the sagittal balance. Uni- or bisegmental fusion provides enough stability. We recommend a dorsal fixation system with low profile after total or subtotal correction of kyphosis, so a ventral compression is achieved. In very young patients, tension wire band or transcorporeal screwing is sufficient. In cases of older children or adolescents, the rudimentary pedicles allow use of an internal fixation device like the Cotrel-Dubousset fixateur interne, which is adjusted to the curve by bending the rods. When the skin is expected to be compromised, instrumentation should be implanted from lateral, so a low profile construct is possible (Fig. 7).

In very young patients the kyphosis should be completely corrected, because of the danger of secondary re-kyphosis. Therefore, the correction should not be delayed, making a correction under reasonable risk of complications almost impossible.

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