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Treatment of scoliosis in intermediate spinal muscular atrophy (SMA type II) in childhood

Key words: *spinal muscular atrophy, spinal surgery, telescope rod*

SUMMARY

Progressive scoliosis with increasing pelvic obliquity in early childhood of patients with SMA type II is a common feature in this disease. Spinal surgery in muscle disorders should be carried out as soon as a progressive curve of more then 20° Cobb and a preserved FVC of 20-30% is proved. In later stages or severe forms of SMA II spinal stabilisation becomes often impossible due to the respiratory insufficiency, the poor general condition and the severity of the scoliosis with marked pelvic obliquity. A special telescope rod was developed in order to enable a lengthening of this instrumentation during growth for children treated in early childhood. In 15 of 20 patients with SMA II in early childhood not satisfactory results after telescope rod implantation were observed. In spite of the telescope technique crankshaft phenomenon appeared and curve progression increased. So then we stopped telescope rod implantation. This instrumentation could be in principle a good therapeutical tool for this indication, but its technical manufacturing has firstly to be improved decisively. For SMA II patients younger than 10 years with progressive scoliosis our therapeutic recommendation is nowadays a corset until the age of 10-12 years followed by definitive surgical correction using other multisegmental instrumentation like the Isola® system.

BACKGROUND

Spinal muscular atrophy (SMA) is a hereditary usually autosomal recessive neuromuscular disease characterised by degeneration of the anterior horn cells of the spinal cord and occasionally of the motor neurons of the cranial nerves 5 to 12. The genetic locus for spinal muscular atrophy has been identified on chromosome 5q and the gene product is the survival motor neuron (SMN) protein. The incidence of SMA is approximately 1 in 15,000 to 1 in 20,000 live births, and the prevalence of the carrier state is 1 in 80 [1]. Proximal and rare distal forms of SMA are observed. There are four different types of proximal SMA (Tab. 1): acute (SMA I), intermediate (SMA II), mild (SMA III) and adult (SMA IV). Type II SMA is usually identified between birth and age 18 months. The children achieve head control, develop the ability to sit but they are unable to stand or walk and require a wheelchair. Contractures of the upper and lower extremities, hip subluxation or dislocation and the development of scoliosis requires orthopaedic treatment in children with SMA II. Additionally apparent chest cage deformities aggravates together with scoliosis the respiratory problems caused by the muscle weakness. The incidence of scoliosis in SMA II is nearly 100%. The scoliosis starts in age of in average 2.1 to 4.3 years and has a progression of approximately 8° Cobb angle yearly [2]. Severe progressive scoliosis already in early childhood in the age of 3 or 4 years can be frequently observed. Long c-formed thoracolumbar curves are most common, seldom thoracic, lumbar or double major curves. Progressive scoliosis (collapsing spine) with pelvic obliquity in SMA II patients (Fig. 1) can not be influenced efficiently by conservative treatment [3]. These scolioses require surgical stabilisation to correct the curve, balance the trunk and to diminish pelvic obliquity in order to improve sitting quality.

Imbalanced proximal muscle weakness leads in a lot of children with SMA II to coxa valga with hip subluxation and dislocation and in some of them to unilateral dislocation [4]. Surgical reconstruction of subluxated or dislocated hip by intertrochanteric and/or pelvic osteotomy in non-ambulatory patients has been described but controversially discussed [5,6].

Tab. 1. Classification and prognosis of different types of autosomal recessive proximal spinal muscular atrophy

Types		Manifestation	Functional grading	Live expectancy
I a	acute	prenatal (30%) to 3-6 months	unable to roll over or sit	< 30 months = 100% < 18 months = 95% < 7 months = 50%
I b	subacute-chronic	like Ia	like Ia	2,5-20 years
II	intermediate-chronic	birth –18 months	sitting	2,5-30 years
III a	mild, retarded motor development	to 3 years	walking	4.-6. decade, usually normal
III b	mild, normal motor development	> 3-18 years	walking	
IV	adult	> 30 years	walking	normal

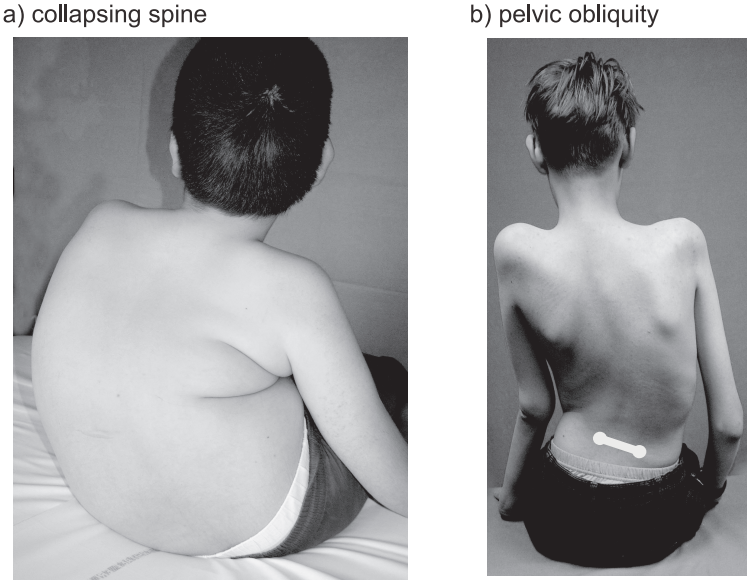

Fig. 1. Main clinical problems of scoliosis in intermediate spinal muscular atrophy: collapsing spine (a) and pelvic obliquity (b)

Summarising hip dislocation or subluxation seems not to be the main causal factor for the pelvic obliquity and sitting discomfort. The decisive factor for the development of pelvic obliquity is the progressive scoliosis. The correct sitting position is most important for non-ambulatory patients and the loss of sitting ability has to be prevented by early surgical stabilisation of scoliosis and pelvis.

CONSERVATIVE AND OPERATIVE TREATMENT

The conservative treatment of scoliosis in SMA II is difficult and ineffective [7]. A sitting support can be used for smaller children without sitting balance with collapsing kypho-scoliosis in order to control sitting position but not to correct spine deformity. Corsets are more useful to make sitting easier in the time be-

fore the operation or when surgery of the spine is impossible, because of high cardiopulmonary risk, poor general condition or high neurological risk like paraplegia after correction of very severe kypho-scoliosis. Corsets can not stop the curve progression. Many children do not tolerate orthoses. Very important is that corsets in SMA II have to distribute the forces over a large area and prevent chest wall deformity while stabilisation sitting position during growth [3].

In spinal surgery of scoliosis in SMA II different instrumentations are used like Luque-, Galveston-, Cotrel-Dubousset- or Isola®-instrumentation. The special advantage of Luque´s segmental spinal stabilisation or its modifications (Galveston or Unit-rod-segmental spinal stabilisation) is the postoperative treatment without corset or plaster. The rods can be adapted to the physiological curves of the spine. The Cotrel-Dubouseset (CD) instrumentation enables a solid foundation in the spine and a better correction of rotation in comparison with Luque´s instrumentation. The most important advantage of Isola® system is the force transmission in three directions. Posterior spinal fusion with multisegmental spinal instrumentation and pelvic fixation is the treatment of choice in patients in SMA II. The instrumentation has to extend from upper thoracic spine (Th2 or 3) to the sacrum or to the pelvis [6,8,9]. Anterior fusion is usually not carried out because of the high surgical risk for injuring the diaphragm as the most important respiratory muscle in these patients.

Extensive pulmonary pre-, peri- and postoperative care is necessary to avoid respiratory complications such as pneumonia and the need for prolonged mechanical ventilation. The preoperative assessment has to include lung function examination and blood gases analysis. Intermediate SMA patients with limited lung function should be adapted to assisted ventilation prior to spinal surgery, since perioperative lung complications can be decisevely reduced. The possible risks of spinal surgery in these very patients can be reduced if special points are followed. The cardiopulmonary situation has to be exactly examined preoperatively. During the operation the prone positioning for surgery has to avoid a direct pressure against the sternum and a heart compression. The anaestesiological regime has to regard the specific demands for patients with muscle disorders. Succinylcholin and halothane are obsolete. Intraoperative monitoring includes EKG, non-ivasive and invasive blood pressure, central venous pressure, arterial blood gases, pulse oxymetrie, capnometrie, patient's core temperature, relaxation and airway pressure. Very important is the correct and consequent regime of volume substitution, dilution during the usually long surgery time with the consequence of partly marked blood loss. Cell-saver has to be used intraoperatively. However, intra- and postoperatively substitution with erythrocyte concentrates and fresh frozen plasma is mostly required. Spinal surgery of intermediate SMA patients is not seldom a hazardous surgery and requires appropriate facilities with experienced surgeons, anaestesiologists and intensive care unit especially in those patients with limited pneumocardial capacity.

Operative stabilisation of the spine in SMA II is indicated in proved progressive curves more then 20° Cobb angle and forced vital capacity (FVC) of at least 20-30% and nowadays the treatment of choice.

PROBLEMS OF SPINAL SURGERY USING THE TELESCOPE ROD

Naumann 1993 developed a special telescope rod for non-fusion surgical spine stabilisation in order to enable a lengthening of this instrumentation during growth for young children primary in Duchenne muscular dystrophy (DMD). A more flexible system with rotational stability and preserved longitudinal mobility compared with Luque-instrumentation was achieved. This system consists of a solid upper rod sliding in a caudal hollow rod, which each connected together on their cranial and caudal end with Labitzke-wires (Fig. 2). The telescope rods were fixed on the spine with polyfile steal cords (Labitzke-wires). To avoid implant failure, corrosion, metal wear and

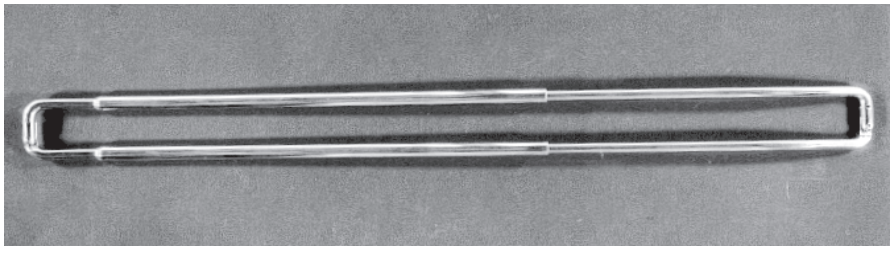

Fig. 2. Original telescope rod

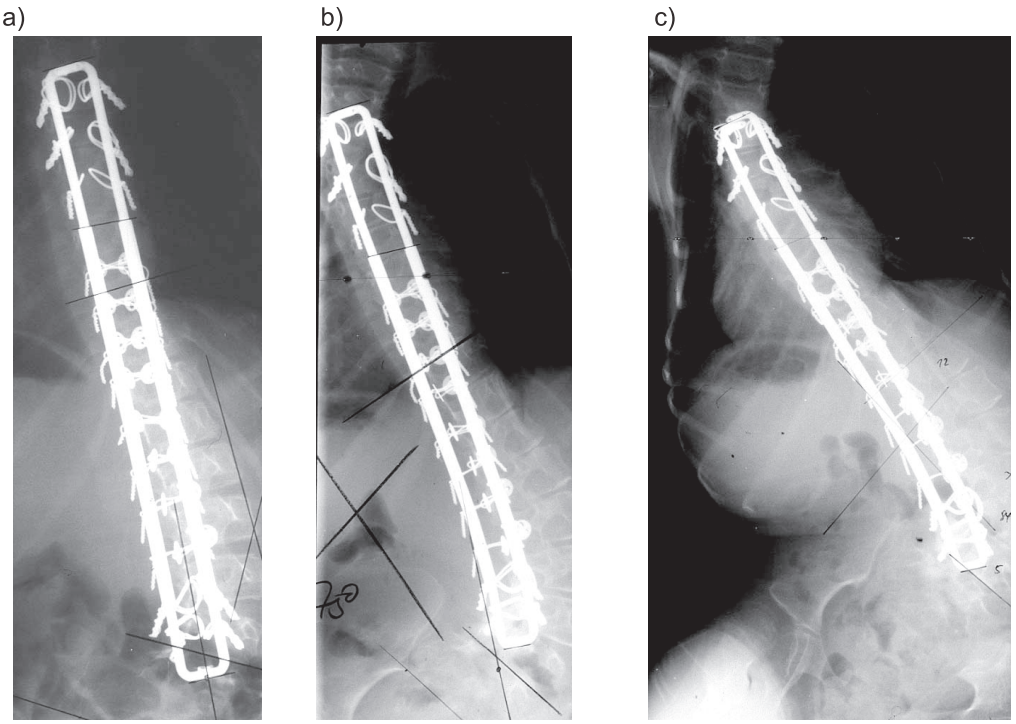

Fig. 3. Crankshaft phenomenon after spinal surgery in SMA II using the telescope rod. a) 0 months postop., Cobb 30°, tilt 29°, b) 14 month postop., Cobb 75°, tilt 56°, c) 22 months postop., Cobb 84°, tilt 52°

foreign body reaction the wires were leaded around plastic rolls. To avoid the osseous resorption and to decrease the risk of wire pull out, the wires were conducted in a metal capsule through the basis of the processus spinosus. This kind of trans-/interspinously anchoring has lowered the risk for neurological complications in the spinal cord and the blood loss.

For the treatment of SMA patients we modified the telescope rod. To avoid disconnecting of the cerclages at the upper and lower end of the rod pair, the ends of them were firstly welded together and later on produced in one piece. For better fixation of the ends of the rods to the spine additional transverse rods were added at the cranial and caudal end. Lateral flattening of the solid rods was done to reduce twisting and canting of the rods against each other.

Heuser (2003) reports about 20 consecutive patients with SMA II operated using telescope rods. The age at spinal surgery was averaged 6.8 years (SD 1.7, range 4.6-10.8 years) and the length of postoperative follow-up was on average 3.6 years (SD 0.7, range 2.5-4.7 years). Pre- and postoperative X-rays, standardised anteroposterior and lateral views were analysed. The development of the Cobb angle was evaluated. Operative correction of the Cobb angle of 46.5° on average (SD 16.8°, range 28°-83°) was achieved (preoperation 62° on average, SD 19.2°, range 25°-90°; postoperation 16° on average, SD 10.1°, range 0°-38°). Clinical analysis showed a mean loss of postoperative correction of 9.6°/year, various significant crankshaft phenomena and an increase of pelvic obliquity (Fig. 3). Because of these clinical results telescope rod implantation in SMA II patients was stopped.

For the biomechanical experiments we used 7 telescope rods and 2 different testing apparatus. The friction forces (sliding and adhesion), in- and out-sliding forces, left/right rod torsion between 0 and 16° in 2° steps, metallic wear and rod deformities were analysed [10]. The experimental investigations showed increased out-sliding forces with increasing torsion angle. A further significant problem was too much metallic wear, and unsatisfactory manufacturing precision with poor surface-quality of the telescope rods.

SURGICAL SPINE STABILISATION USING THE ISOLA® INSTRUMENTATION

Isola®-system (AcroMed) is a spinal instrumentation based on the „3 dimensions – 6 motions" (3D--6M) principle. For spinal stabilisation a combination of hooks, sublaminar wires and pedicle screws are

used and allows a good segmental correction and stability. Our technique: The instrumentation includes transverse process-lamina hook claws at Th_2 and Th_4 or Th_3 and Th_5. We use double Luque-wires from Th_5 (Th_6) down to Th_{12}, pedicle screws in L_1-S_1, transverse connectors at Th_3 (Th_4) and L_3. Bony fusion with bank bone from L_1 to sacrum was used in the first few cases. Later on we perform autologous bone grafting only between L_5 and S_1 using the resected processus spinosi. Furthermore a decortisation of the vertebra joints is performed at the end of the operation. Immediately after surgery the instrumentation is solid enough for sitting and transfers without corset. Mobilisation in wheelchair occurs as rapidly as possible, mostly within 6-8 days postoperatively.

From January 1999 until August 2004 17 SMA II patients have been operated for progressive scoliosis using the Isola®-system. The average age et spinal surgery was 12.9 years (SD 2.9, range 9.4-22.7 years). Operative correction of the Cobb angle of 41° on average (SD 17.7°, range 2°-82°) was achieved (preoperation 82.7° on average, SD 17°, range 54°-120°; postoperation 44.4° on average, SD 19.4°, range 17°-88°).

CONCLUSION

Near 100% of SMA II patients develop severe progressive scoliosis with increasing pelvic obliquity already in early childhood in the age of 3 or 4 years. The development of scoliosis of growing spine in early childhood is the most significant problem of orthopaedic treatment of these children. Conservative treatment of scoliosis in SMA II is ineffective. The progressive scoliosis with pelvic obliquity in young patients with SMA II have to be surgical treated by spinal stabilisation. The telescope rod could be in principle a good therapeutical approach for stabilisation of growing spine, but its technical manufacturing has firstly to be improved decisively. Reduction of friction forces (sliding and adhesion), reduction of metallic wear, and better technical precision of production are necessary. Our clinical investigation has shown not satisfactory telescoping of telescope rods in a large percentage of patients. Unsatisfactory precision of technical manufacturing and poor surface--quality of telescope rods could be proved by our biomechanical experiments as causal factors. For SMA II patients younger than 10 years with progressive scoliosis our therapeutic recommendation is nowadays a corset until the age of 10-12 years and then the definitive surgical correction using other multisegmental instrumentation like the Isola® system. From the technical point of view we recommend the instrumentation from Th_2 or Th_3 down to S_1 combined with bony fusion in order to improve pelvic obliquity for better sitting balance. Early spinal surgery, however, improves both sitting comfort of the patients and quality of life for the patients and their families. New instrumentation for spinal stabilisation in the early childhood have to be developed for these very patients.

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