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GRAND ROUNDS

Combined selective dorsal rhizotomy and scoliosis correction procedure in patients with cerebral palsy

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Abstract

Intrathecal baclofen (ITB) therapy for spasticity has been suggested to accelerate the development of scoliosis. We present the case of a 17-year-old female patient with cerebral palsy who had ITB therapy from the age of 11 years. During this period, she developed a severe scoliosis measuring 86° from T11 to L4, with pain due to costo-pelvic impingement. Her baclofen pump had reached its end of life and required replacement if ITB therapy was to continue. This coincided with plans for scoliosis corrective surgery.
Methods We performed scoliosis correction along with removal of baclofen pump and selective dorsal rhizotomy (SDR), as a single combined procedure. SDR was performed instead of ITB pump replacement for management of spasticity.
Results Following surgery, scoliosis improved to 24°. At 6 month follow-up, there was significant improvement in spasticity and quality of life.
Conclusions This report illustrates the feasibility of a combined procedure to correct scoliosis and manage spasticity with SDR. We present the case details, our management and review of the published literature regarding the factors influencing treatment of scoliosis and spasticity.

Keywords Spasticity · Scoliosis · Cerebral palsy · Baclofen · Selective dorsal rhizotomy

Introduction

Use of intrathecal baclofen (ITB) therapy for management of spasticity has been associated with development of, and accelerated progression of neuromuscular scoliosis [1, 2]. We present a case of a patient who developed a severe lumbar scoliotic curve during 6 years of ITB therapy. This report outlines our management of coexisting spasticity and scoliosis.

Case details

Our patient was diagnosed with cerebral palsy (CP), with spastic quadriparesis and intellectual delay. She was first referred to our unit at the age of 11 years for the management of her spasticity with GMFCS (Gross Motor Function Classification System) grade 5 (severely limited self-mobility and little head or trunk control). Feeding was

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assisted via gastrostomy. There were no features of dystonia. She had significant flexion contractures of all of her limbs and bilateral subluxation of ankles and wrists, as identified from assessment under anaesthesia.

We managed her spasticity with ITB therapy from the age of 11 years, after positive results from single bolus test dose of intrathecal baclofen. The baclofen pump implanted was a Medtronic Synchromed® II drug delivery system (Medtronic, Minneapolis, USA). Surgery had been uneventful and there were no pump related complications for the duration of ITB treatment.

Initial ITB dose delivery was set to 60 micrograms per 24 h and her carers reported an improvement in her spasticity. Since then her baclofen requirements increased and at the age of 17 she was requiring 460 micrograms per 24 h and pump refill was required every 2 months. The GMFCS grade remained 5 and Ashworth score was 3 throughout all of her limbs.

Spine radiographs at the age of 11 showed an almost straight spine. During the subsequent 6 years of ITB treatment, we noted a progressive scoliosis. At the age of 15 she had a lumbar curve with a Cobb angle of 40° from T11 to L4. By the age of 17 years this had progressed rapidly to a measurement of 86° from T11 to L4 (Fig. 1). Her spinal deformity seemed to be causing significant pain from costo-pelvic impingement. Nursing care had also become more difficult.

The need for surgical correction of her spinal deformity coincided with the need for surgery to address spasticity management, as her baclofen pump was reaching the end of its battery life. Given the difficulties posed by regular baclofen refills and the increasing ITB dose requirements her parents agreed to selective dorsal rhizotomy (SDR), as an alternative to replacing the baclofen pump and continuing ITB therapy. We, therefore, planned to remove the baclofen pump and perform SDR and scoliosis correction as a single procedure.

Our preoperative workup included MRI of the brain and whole spine. This excluded intraspinal pathology. The level of the conus was at L1. Brain MRI showed mature cerebral white matter volume loss in keeping with periventricular leukomalacia, and no other pathology. Other investigations performed included urological assessment to evaluate baseline function. The delivery of baclofen was weaned in the run up to surgery to avoid ITB withdrawal syndrome.

Diagnostic imaging

Surgical procedure

We performed a staged procedure under a single general anaesthesia (GA). Traction radiograph showed a fairly rigid curve, which was only reducible to 49° from T11 to L4.

With the patient placed supine, we performed the first surgical procedure, which was removal of baclofen pump and catheter though an incision over the abdominal scar. The subcutaneous tunnel that had accommodated the intrathecal catheter was closed, to prevent leakage of cerebrospinal fluid into the abdominal site.

The patient was placed prone on a Montreal mattress and incision was made to allow exposure of the spine. We proceeded with segmental pedicle screw placement from T2 to L5, and bilateral facetectomy at each of the lumbar levels. Iliac screws were also placed for pelvic fixation. We

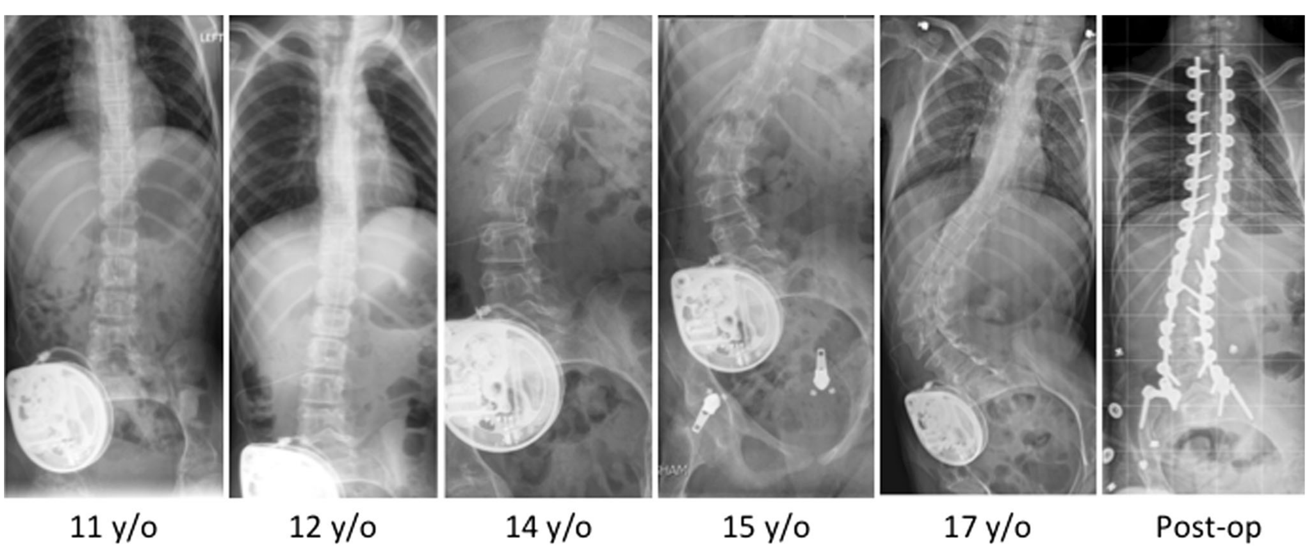

Fig. 1 Spine radiographs showing a progressive scoliosis from the age of 11 years to the age of 17 years, resulting in a curve of 86° from T11 to L4. The final sitting radiograph was performed at 6 months following surgery and shows correction of scoliosis to 24° from T11 to L4

were able to achieve significant derotation without performing additional osteotomies. Spinal cord monitoring was performed with motor evoked potentials (MEPs) and somatosensory evoked potentials (SEP).

SDR was subsequently performed via a T12 to L1 laminectomy, followed by a midline durotomy. The dorsal roots were identified by anatomical position. L1 nerve roots were identified by their exit under the L1 pedicle. S1 nerve roots were easily identified by their considerably larger size compared to other roots. The rootlets forming each nerve were gently separated using micro dissectors (Rhoton™ #6) and we proceeded to cut approximately 2/3 of the rootlets at each nerve root from L2 to S1 (Fig. 2).

After dural closure, rods were contoured and fixed to the pedicle screws, allowing correction of the scoliosis. We initially placed a temporary rod on the convexity side to partially reduce the deformity before placing the permanent rods. This allowed us to achieve a greater degree to derotation and deformity correction. Autograft was used to encourage fusion.

Total operating time was 520 min, following which the patient was transferred to paediatric ICU and kept ventilated overnight. Intra-operative blood loss had been 1000 ml. Cell salvage had been used and we were able to replace 400 ml during the procedure. Post-operative blood transfusion was not necessary.

Once sedation was stopped the patient woke fairly promptly and was discharged from hospital 4 days after surgery. Physiotherapy was started in hospital and continued in the outpatient setting. There were no post-operative complications.

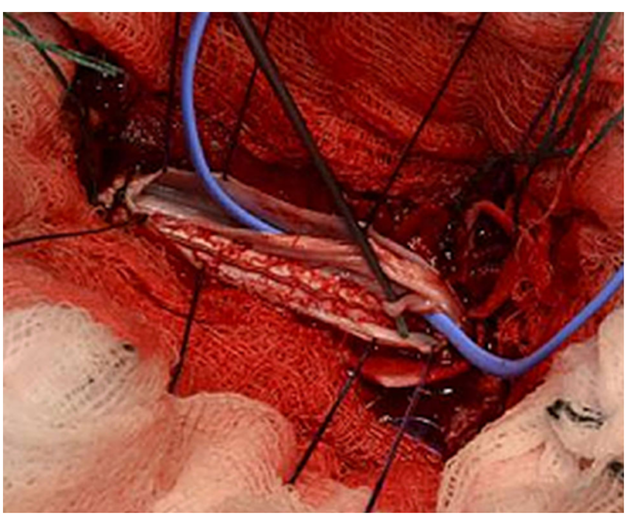

Fig. 2 Intraoperative photograph during SDR stage. Dorsal roots from L2 to S1 were identified and a silicone loop was used to isolate them from ventral roots. Micro dissectors (showing the S1 nerve in this figure) were used to separate the rootlets comprising each nerve. 2/3 of the rootlets at each nerve root we subsequently cut

Post-operative follow-up

Post-peratively we achieved scoliosis correction from 86° to 24° (T11 to L4). At 6-month review, these findings were static and there were no hardware related problems. Ashworth scores in the lower limbs improved from 3 pre-op to 0, as did Ashworth scores in the upper limbs. There was no loss of motor power and bladder function was unchanged. CPCHILD™ questionnaire showed quality of life improvement with total scores of 54.4 % pre-operatively increasing to 64.3 % at 6-month follow-up. Areas of most significant improvement were in the domains of Personal Care and Comfort.

Discussion

Treatment of scoliosis

Cerebral palsy (CP) has an estimated incidence of 1–2 per 1000 live births [3]. Of the neuromuscular disorders CP has the highest association with scoliosis; incidence is estimated to range from 21 to 76 % in the CP population [4]. More severe forms of CP are associated with a higher incidence of scoliosis [5].

Although the use of intrathecal baclofen (ITB) is effective in the management of spasticity and dystonia, there has been considerable concern regarding acceleration in scoliosis progression with rates increasing to 18.4° per year [1, 2]. This contrasts with the observed curve progression rates in CP patients without ITB therapy, which is in the region of 4.4° per year [2].

The Heuter–Volkmann law applied to scoliosis leads to the assumption that muscle imbalance produces abnormal loading of the spine and leads to asymmetrical vertebral growth, therefore, resulting in progressive scoliosis. It is possible that reducing muscle tone with ITB treatment accelerates development of a progressive scoliosis by worsening the asymmetrical vertebral load.

Scoliotic curves of less than 30°, where there is no evidence of progression or loss of function, can be managed with conservative measures [6]. However, in curves greater than 50°, or where there is evidence of progression or loss of function, surgical correction should be considered.

Surgical correction of scoliosis in patients with CP is associated with high patient and carer satisfaction rates. Comstock et al. [7]. found an 85 % satisfaction rate, with parents and carers reporting a positive impact on the patient's sitting ability, comfort, ease of nursing care and physical appearance.

The reported incidence of postoperative complications, including wound infection and failure of instrumentation, varies from 18 to 68 % [7]. Risk of complications is highest when curves are 70° or greater and neurologic involvement is severe [6].

Our recommendation is for instrumentation from T2 to pelvis. The incidence of proximal curve progression, and proximal junctional kyphosis is higher if the proximal level of instrumentation does not extend to T2 [8], in cases where the patient lacks good head control. Pelvic obliquity may progress if fusion is not extended to include the pelvis [9, 10].

Management of spasticity

Selective dorsal rhizotomy (SDR) and intrathecal baclofen (ITB) are both proven treatment modalities to alleviate spasticity for children with cerebral palsy [11–13]. Traditionally SDR has been reserved for ambulant spastic diplegic patients (GMFCS 2–3) to improve gait, whilst ITB has been used in GMFCS 4–5 cerebral palsy patients to reduce spasticity and facilitate nursing care.

There are limited studies evaluating outcome of SDR in GMFCS 4–5 patients. Kan et al. [14]. directly compared results of SDR and ITB implantation in patients with severe spasticity (GMFCS 4 and 5). SDR emerged as a superior treatment, with significant improvements in Ashworth scores, passive range of motion in the lower extremities and change in GMFCS grade, compared with ITB. In addition, a recent review of all spasticity interventions confirmed SDR as an effective and superior treatment over ITB [13]. Removing the ITB system and performing SDR eliminates the need for regular baclofen pump refills and also catheter and pump related complications, which are in the region of 15 % [15].

Improvements in quality of life and functional status following SDR have been reported previously [16]. Gigante et al. [17] reported that more than 90 % of patients with spastic quadriplegia undergoing SDR also had a reduction in upper limbs tone and half of them reported functional improvement. In addition, Craft et al. [18]. reported cognitive improvement following SDR and they attributed this to improvement in mood, reduced physical discomfort, reduced therapeutic intervention, and possible cortical effects of SDR.

The daily dose of ITB required for management of spasticity typically increases over time. Development of baclofen tolerance is a major concern with ITB therapy [19]. It occurs due to down-regulation of GABAb receptors in the spinal cord following chronic baclofen infusion [20], thereby leading to the loss of efficacy of baclofen.

We have performed over 300 ITB system implants for spasticity in children over the past 15 years, and have performed SDR instead of replacing ITB systems at the end of pump life, in 14 patients with GMFCS grade 4 and 5. Our findings are the same as those reported by Kan et al. [14] with improvement in spasticity, quality of life and level of nursing needs in each of these patients following SDR compared to when they had optimal ITB therapy. These effects are maintained at 2 years follow-up. In addition, Dudley et al. [12]. showed that improvements in spasticity following SDR are maintained at 15-year follow-up. Some of the functional benefits, however, are lost over the same follow-up period but remain significantly above levels prior to SDR. We offer SDR to patients regardless of GMFC grade, although in patients with GMFCS grade 4 and 5 our primary aim is to manage spasticity rather than improve function.

A combined procedure can be performed irrespective of whether the patient has previously had ITB therapy; although in our experience SDR is technically more challenging where long-term ITB therapy has been used, as this leads to arachnoid fibrosis.

Early SDR techniques involved laminectomy from L1 to S1. However, we use a technique similar to that described by Park and Johnston [21], which utilises a single level laminectomy over the conus medullaris. Park and Johnston [21] also described the use of an interlaminar dissection and intraoperative ultrasound scan to confirm the location of the conus prior to laminectomy and durotomy. We find this step unnecessary as we perform MRI scans routinely in our pre-operative assessment, and from this we can identify the level of the conus.

Although we do use intraoperative neurophysiological monitoring, we are not reliant on this when the anatomy is easily delineated. Dorsal roots can be differentiated from ventral roots by their anatomical position. The L1 nerve roots are identified as they exit the neural foramina and the S1 nerve roots are typically large and followed by S2 nerve roots, which are very small. Neurophysiological assessment is useful in cases where there is uncertainty. We do not use neurophysiological stimulation to select the rootlets to cut, as several studies have shown intraoperative electromyography to be inconsistent when repeated and unreliable. Moreover, Steinbok et al. [22] found no difference in outcome between patients undergoing SDR with neurophysiological guidance and those without.

Combined procedure to address scoliosis and spasticity

Our case demonstrates that a combined procedure is feasible and we suggest that this should be considered in cases where there is significant spasticity and scoliosis that requires surgical correction. The advantages of a combined procedure include avoidance of several general anaesthetics, a shorter overall operating time and shorter inpatient stay. It also avoids delay in treatment whilst waiting for the patient to recover from one procedure before undergoing the next.

Conclusion

In patients with spasticity and significant scoliosis, a combined procedure with posterior instrumented correction and SDR should be considered. Our case demonstrates that this is feasible.

There is evidence to suggest that scoliosis is likely to progress in patients with CP; and this progression is faster in the presence of ITB therapy. SDR with scoliosis correction should be considered in cases with or without pre-existing ITB therapy. SDR has been shown to provide better spasticity control and improve quality of life. This is the case for all GMFCS grades, even in severe spastic quadriparesis.

Our patient showed improvement in both upper and lower limb spasticity. Outpatient follow-up schedule for both scoliosis clinic and functional neurosurgery clinic are 6 weeks post surgery, 3 months, 6 months and then every 6 months until 2 years is reached. Subsequently follow-up is on a yearly basis until 5 years is reached, at which point the patient is discharged. We expect the improvement seen in this patient to be maintained at long-term follow-up based on the published literature [12] and from our previous clinical experience in other patients.

Compliance with ethical standards

Conflict of interest None.

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