Freehand correction of scoliosis in Rett's syndrome

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Abstract We present the case of an 18-year-old girl with a scoliosis (long C-shaped curve) in association with an atypical Rett's syndrome. In order to attain a full correction and to provide her with adequate posture and sitting balance, segmental instrumentation was utilised from the high thoracic spine to the pelvis. We describe the procedure including the relevant pre-operative, intra-operative and post-operative imaging.

Keywords Scoliosis · Rett's syndrome · Segmental screw · Freehand

Case presentation

An 18-year-old female with both an atypical Rett's syndrome and Lennox–Gastaut syndrome [4–7] presented to our unit with a progressive scoliosis, which behaved much like a neuromuscular long C-shaped curve.

Clinically, the patient had significant developmental delay and was wheelchair bound. There was limited function in either lower limb. She was incontinent of urine and faeces and a gastrostomy was in place due to variable and sometimes inadequate oral intake. Severe epilepsy, due

to the Lennox–Gastaut syndrome, was managed by a vagal nerve stimulator (visible on the plain radiographs), which was controlled by an external magnet. In addition, the epilepsy was also managed more conventionally, using traditional pharmacological means. Despite both these interventions, our patient suffered from frequent, daily seizures, which were variable in nature.

From a clinical point of view, surgery was deemed necessary due to an increasing loss of sitting balance and the potential for respiratory compromise. Annual monitoring of the curve deemed the scoliosis to be progressive and this provided a further mandate for surgical intervention. Pre-operative radiographs (Fig. 1) revealed a long C-shaped curve. In addition to whole-spine (sitting) radiographs, a whole-spine MRI was obtained which revealed no craniocervical or intraspinal abnormality. Pre-operative radiographs were used to quantify the Cobb angle as approximately 50°.

The patient's family was thoroughly counselled and consented to the proposed surgery. Proposed benefits as well as risks were discussed and documented.

Procedure

Prior to the procedure the operating table and Montreal mattress were adjusted to accommodate our patient's relatively light frame.

Pre-operative planning of the scoliosis surgery was undertaken, using a whole upright spine radiograph

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Fig. 1 Pre-operative and post-operative (sitting) whole-spine radiographs, demonstrating a 50° long C-shaped curve with excellent correction of the deformity in both sagittal and coronal planes

(sitting). The radiograph was marked to allow identification of both the convex and concave pedicles to aid the freehand technique. A red dot was applied to the convex pedicles and a black dot to the concave pedicles (Fig. 2). The red dots convey to the surgeon that the pedicle is visible and that a more vertical trajectory than normal is required. Conversely, the black dot requires the placement of the pedicle screw in a more horizontal trajectory. This facilitates the correct placement of pedicle screws in the deformed spine with greater accuracy, when using the freehand technique [1, 3].

After general anaesthesia was administered, spinal cord monitoring electrodes were placed and secured to allow monitoring of somato-sensory evoked potentials. Motor-evoked potentials were not utilised for fear of aggravating our patient's already severe epilepsy.

The patient was positioned prone and pressure areas identified and protected. After sterile draping, a midline posterior longitudinal incision, extending from T1 to the sacrum, was made. Monopolar diathermy was utilised to perform a subperiosteal dissection of the posterior spinal muscles from the underlying bone, with exposure along the lamina, and laterally to the transverse processes. The subperiosteal dissection took place from a caudal to a cephalad direction at each level, along the line of the lamina. This is important in the elevation of the multifidus muscle, which attaches to the posterior elements in an oblique fashion from a caudal to cephalad direction.

The transverse processes were identified by direct palpation at each level. Dissection was carried out until

exposure of both the superior and inferior margins of the transverse processes was achieved. Care was taken to prevent exposure anteriorly, in order to avoid damage to the exiting nerve roots. Haemostasis was achieved by bipolar diathermy. Having exposed the spinous processes, laminae and transverse processes, self-retaining retractors were applied and haemostasis was once again ensured. Attention turned to appropriate preparation of the pedicles with identification of the appropriate pedicle entry site: in the thoracic spine this is at the junction between the superior border of the transverse process and the lamina; and in the lumbar region this at the distal pole of the superior facet.

It is important to appreciate, however, that often the pedicles are dysplastic, particularly on the concave side, and they may require gentle expansion using the reamer. This is usually feasible if sufficient time is allowed to expand the walls of the dysplastic pedicle. However, if this is deemed to be impossible, extra-pedicular screws should be placed. Pedicle screws were then inserted along the path [2, 8, 9].

After the screws were inserted, the length of the rods were determined and cut to size. A thoracic kyphosis and lumbar lordosis were carefully contoured and the distal aspect of the rod was adjusted to accommodate the pelvic fixation.

In order to obtain pelvic fixation, adequate exposure of both pelvic brims is essential. A handheld retractor and diathermy were used to mobilise a thick flap to expose both pelvic brims. A triangular section from the posterior superior iliac crest was removed in order to allow the placement of a pelvic screw level with the S1

Fig. 2 Pre-operative planning for freehand technique (*red dots* convex pedicles, *black dots* concave pedicles); instrumentation levels and screw sizes

screw. Care was taken to ensure that neither the inner nor outer tables of the pelvis were breached. A polyaxial screw was inserted into the pre-prepared tract on both sides.

The rods were placed, and by using two-rod holders, correction of the curve was achieved by way of a derotation manoeuvre in order to correct the deformity and restore the thoracic kyphosis and the lumbar lordosis. The rod was then secured in position by final tightening of all screw caps. The second rod was introduced and secured in the same manner. The pelvic screws were attached to the rods using appropriate cross-links.

The final step of the procedure was decortication. In addition to autograft, tricalcium phosphate synthetic bone graft strips, perfused with the patient's own blood, were applied. The surgical incision was closed. A chest radiograph was obtained in the operating theatre to exclude a pneumothorax.

There were no intra-operative complications and no loss of spinal cord monitoring throughout the procedure. The total blood loss was approximately 600 ml and the operative duration was approximately three-and-a-half hours.

Outcome and follow-up

Our patient was hospitalised for 7 days. She was noted to be rising to a seated position after 3 days with greater ease, and her family were pleased with the improved sitting posture and stable sitting balance.

There was neither deterioration nor improvement in lower limb, bladder or bowel function. The patient was discharged when her wound was deemed to be dry and healed.

Post-operative radiographs revealed a full correction of the scoliotic deformity with good restoration of the thoracic kyphosis and lumbar lordosis.

Conflict of interest None.

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