CASE REPORT

Posterior occipitocervical fixation under skull-femoral traction for the treatment of basilar impression in a child with Klippel–Feil syndrome

We present the case of a 15-year-old boy with symptoms due to Klippel–Feil syndrome. Radiographs and CT scans demonstrated basilar impression, occipitalisation of C1 and fusion of C2/C3. MRI showed ventral compression of the medullocervical junction. Skull traction was undertaken pre-operatively to determine whether the basilar impression could be safely reduced. During traction, the C3/C4 junction migrated 12 mm caudally and spasticity resolved. Peri-operative skull-femoral traction enabled posterior occipitocervical fixation without decompression. Following surgery, cervical alignment was restored and spasticity remained absent. One year after surgery he was not limited in his activities. The surgical strategy for patients with basilar impression and congenital anomalies remains controversial. The anterior approach with decompression is often recommended for patients with ventral compression of the medullocervical region, but such procedures are technically demanding and carry a significant risk of complications. Our surgical strategy was an alternative solution. Prior to a posterior cervical fixation, without decompression, skull traction was used to confirm that the deformity was reducible and effective in resolving associated myelopathy.

The common feature of Klippel–Feil syndrome (KFS), a rare congenital condition first described a century ago, is incomplete segmentation of two or more cervical vertebrae. The clinical features include the triad of a low posterior hairline and a short neck with limited movement. However, fewer than half of the patients with KFS exhibit all three of these traits. The syndrome often presents with associated disorders including deafness, facial anomalies, scoliosis and cardiovascular and genitourinary abnormalities. As patients with KFS are often asymptomatic, the incidence is unknown; estimates suggest an incidence of between 1 in 2100 and 1 in 42 400 births.

Basilar impression with occipitalisation of the atlas and spontaneous fusion of C2/C3 is sometimes seen in these patients. The odontoid process may migrate upwards causing compression of the medulla oblongata and spinal cord. Symptoms include headache, upper motor neurone lesions and lesions of the cranial nerves. Surgical treatment, usually via an anterior approach, involves decompression, sometimes with division of the maxilla. However, anterior procedures are technically demanding and carry the risk of complications, including infection, swelling of the tongue and persistent hoarseness.

We describe the management of a skeletally immature patient with KFS who underwent reduction and posterior stabilisation, without anterior decompressive surgery.

Case report

A 15-year-old boy was referred with myelopathy. There was a long history of limitation of movement of the neck. Sixteen months before presentation he became unable to throw a ball, and gradually became unable to perform fine movements with his right hand.

Physical examination demonstrated a short neck, a low hairline and restricted movement of the neck. There was hyperreflexia in the lower limbs, positive ankle clonus and an extensor plantar reflex. The grip-and-release test (10-second test) was 18 times/10 s and 19 times/10 s in the right and left hands, respectively. Muscle power measured by Manual Muscle Testing and light touch sensation was normal in the upper and lower limbs except for some wasting of the thenar muscles in both hands.

Radiological examination demonstrated craniovertebral abnormalities, scoliosis in the cervicothoracic region and fusion of C2/C3 (Fig. 1a). The distance from the C3/C4 level to McGregor’s line (drawn from the postero-superior tip of the hard palate to the caudal...
base of the occiput)\(^\text{16}\) was 19 mm. Dynamic lateral radiographs demonstrated subaxial subluxation at C5/C6. CT scans showed basilar impression (which could not be accurately quantified due to the abnormalities specific to this case) and assimilation of the occiput with the atlas (\(C1\)) (occipitalisation). CT angiograms showed anomalies of the vertebral artery bilaterally. The left vertebral artery was wider than the right and followed an abnormal course between the foramen transversarium of \(C2\) and the foramen magnum. Appearances suggested a significant risk of vascular injury with screw fixation of \(C1\), \(C2\) or \(C3\) vertebrae. The \(C4\) pedicles, however, appeared normal. MRI demonstrated ventral compression of the medulla oblongata and the spinal cord. The cervicomedullary angle was 147° (the normal range of which has been reported to be between 139° and 175.5°\(^\text{17}\)) (Fig. 2a) and axial MRI showed that the cervical spinal cord was not compressed posteriorly.

Pre-operatively we performed skull traction, with the patient awake, to confirm the potential for reduction. Using a halo ring (GSS Head Grip Arc system; Tanaka Medical Instruments Co. Ltd, Tokyo, Japan), traction was begun with 5 kg and increased to 7 kg in 1 kg increments every four hours. With traction, the deep tendon reflexes in the lower limbs returned to normal and ankle clonus disappeared. Lateral radiographs taken during traction showed partial reduction vertically. The distance from McGregor’s line to the \(C3/C4\) level was 31 mm, suggesting a 12 mm translation of the cervical spine caudally (Fig. 1b). Posterior occipitocervical fixation was then performed under traction, without decompression.

With the patient under general anaesthesia in a prone position on a GSS Head Grip Arc system (Tanaka Medical Instruments Co. Ltd) and a Hall’s four-point frame, we used an image intensifier to confirm that halo-femoral traction with 10 kg provided alignment similar to that obtained with pre-operative traction. A posterior midline longitudinal skin incision was made from the external occipital protuberance to the spinous process of \(C6\). The spine was exposed bilaterally to the lateral margin of the facets, and screws were inserted into the \(C4\) pedicles. Cortical screws were inserted into the occiput and fixation was obtained to \(C4\), using an RRS Loop Spinal System (Robert Reid Inc., Tokyo, Japan). After decortication, bone graft harvested from the ilium was applied and fixed in rods with ultrahigh molecular-weight polyethylene cables (Nesplon; Alfa Inc., Osaka, Japan) in order to keep the graft immobilised. No abnormal waveforms were detected on intraoperative spinal monitoring.

Immediately post-operatively the deep tendon reflexes were normal. A Philadelphia cervical collar (Össur, Reykjavik, Iceland) was used for four months. One year post-operatively he had resumed normal activities. Radiographs confirmed union of the bone graft from the occiput to \(C4\). (Fig. 1c). The distance from McGregor’s line to \(C3/\) \(C4\) remained at 31 mm. MRI showed vertical partial reduction of the odontoid process, which still remained intracranially, with a cervicomedullary angle of 163° (Fig. 2b). As an alternative to a clivoaxial angle we measured the angle between the line connecting the posterosuperior tip of the sella turcica and the cranial tip of \(C2\), and the posterior wall of the \(C2/C3\) vertebral body on MRI. This angle increased from 113° pre-operatively to 137° post-operatively. The distance between the posterior tip of clivus and the \(C3/C4\) level also increased from 40 mm pre-operatively to 47 mm post-operatively.

**Discussion**

KFS is characterised by fusion of two or more cervical vertebrae with or without associated spinal and general abnor-
operative distraction manoeuvre between the occiput and the C1 is strongly associated with instability and myelopathy. Some have described an intraoperative distraction manoeuvre between the occiput and the vertebral artery and sudden death. The use of skull traction in the management of craniovertebral abnormalities has also been described. Salunke et al managed irreducible atlantoaxial dislocation by transoral decompression followed by a posterior fusion, once reduction had been achieved with Crutchfield cervical traction. Simsek et al reported successful correction of basilar impression by posterior decompression and fusion after four weeks of halo traction. Although patients in their series had less basilar impression than our patient (compared on plain lateral radiographs), the authors chose to perform a decompression. According to an algorithm for the management of basilar impression proposed by Goel, Shah and Rajan, the irreducibility of basilar impression in our case required either atlantoaxial distraction with fixation or posterior fixation following transoral decompression. We confirmed that spasticity of the lower limbs resolved during pre-operative skull traction, although complete reduction of basilar impression was not obtained, and could also not be accurately measured. In addition, we determined that posterior decompression was unnecessary because an axial MRI scan showed sufficient subarachnoid space behind the spinal cord.

Surgery for basilar impression in patients with KFS remains challenging and a standard surgical strategy does not exist. Our management involved skull traction in an awake patient three months before surgery to confirm the reduction of basilar impression and assess improvement in neurological function, followed by occipitocervical fixation augmented by bone grafting, under skull-femoral traction. This treatment strategy gave a satisfactory outcome in this patient.

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References


Fig. 2a
MRI scans a) pre-operatively, showing ventral compression of the medulla oblongata (white arrow), and b) at three months post-operatively, showing diminished ventral compression.

Fig. 2b
The angle between a line connecting the posteroinferior tip of the sella turcica to the cranial tip of C2, and the line of the posterior wall of the C2/C3 vertebral body (solid white line) had increased after surgery, as did the distance between the posteroinferior tip of the clivus and the C3/C4 level (dotted white line).
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Myelopathy hand: new clinical signs of cervical cord damage.

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