

## **High cervical fixed severe kyphosis in adult cases with Down syndrome**

**Background:** Although the number of adult patients with Down syndrome has increased, there are no coherent reports on the surgical treatment of its high cervical kyphosis. We report our clinical results of surgical treatment for this intractable deformity.

**Materials and Methods:** Consecutive six adult Down syndrome patients with severe high cervical kyphosis who were operated on from 2014 to 2020. Age at surgery: 20-48 years (avr. 28.8 years), common skeletal abnormality: os odontoideum and steep slope of the C1/2 facet joint. Preoperative symptoms: difficulty in raising the upper limbs, non-ambulatory or severe gait disturbance due to myelopathy/ataxia. All cases underwent occipito-cervical corrective fusion. Post-operative follow-up period: 24 -60 months (avr. 42.3 months). Perioperative complications, bony union, and correction status were examined. JOA scoring for neurologic evaluation could not be utilized because of their poor communication.

**Results:** Solid bony union was obtained in all cases. Average C0-C2 correction angle was 27 degrees. Four non-ambulatory patients became to walk independently postoperatively. Five patients who could not raise their upper extremities were able to raise them after surgery. Postoperative major complications; DJK developed in two patients and required caudal extension of fixation. Allodynia and infection occurred in one patient, respectively.

**Discussion and Conclusions:** Although these surgeries are technically demanding, even non-ambulatory patients can be expected to improve sufficiently. Intra-articular dissection in C1/2 facet joint was the key procedure for realignment in most cases. Preoperative data on sagittal alignment and balance were obtained only from sitting radiographs due to severe paralysis, making it difficult to compare the results with already-known sagittal morphologic studies using standing radiographs. Further discussion will be required on this issue.

**Figure:** Typical preoperative radiograph

