Clinical Evaluation of Surgical Treatment for the Upper Cervical Instability Associated with Klippel-Feil Syndrome

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Abstract

**Purpose:** To evaluate the clinical efficacy of preoperative and postoperative treatment of patients with Klippel-Feil syndrome (KFS).

**Methods:** 8 patients with KFS with cervical instability were selected as the subjects during April 2008 to June 2015, including 3 males and 5 females, who aged 25 - 45 years. These 8 patients were accompanied by cervical spine instability of C2-3 congenital fusion, surgical treatment, postoperative rehabilitation training and health education, regular review of imaging tests, physical examination, as well as assessment by Japanese Orthopaedic Association (JOA) and American Spinal Injury Association (ASIA) scoring systems.

**Results:** The 8 patients were successfully followed up for surgery. Statistical analysis results of all patients with lateral cervical spine, no internal fixation failure, were well graft fusion, in which 8 patients were no incontinence, no obvious spinal cord, nerve root, and vertebral artery injury. However, only 1 patient after 5 months showed recurrence of atlantoaxial dislocation, which led to the second hospitalization. The final postoperative score of JOA was significantly increased compared with the preoperative score, as well as the ASIA sensory score and exercise score. In one patient, a three dimensional (3D) printing guide plate was used to assist pedicle screw placement, which was accurate and safe.

**Conclusion:** Surgical treatment of KFS patients with cervical spine instability is still risky, however, surgical treatment can alleviate or cure the postoperative symptoms. Here, 8 patients were satisfied with the results of surgical treatment, therefore, the surgical treatment with cervical instability in patients with KFS is a highly effective treatment strategy.

**Keywords:** KFS; Upper Cervical Spine; Surgical Treatment; 3D Printing Guide Plate; Japanese Orthopaedic Association (JOA) Score

Introduction

To evaluate the clinical efficacy of preoperative and postoperative treatment of patients with Klippel-Feil syndrome (KFS).

A Typical Case

The patient, xiang × ×, female, 25 years old, "neck discomfort with headache for more than 5 months"; the patient had neck discomfort without obvious inducement before 5 months, the neck activity was slightly restricted, there is a sound during the activity, the right side is migraine, and sometimes it is radiated to the right eye. The patient has not paid attention to it, and the symptoms of head and neck discomfort are aggravated. MRI of the cervical vertebrae is showed that the axial dentate process moved backward, the medulla oblongata and cervical curvature increased, and the signal in the medulla was less homogeneous. The patient was admitted to hospital with "cranial depression". Specialist examination: cervical tenderness (+), especially around the 2 spinous processes of the neck. the right neck and
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Facial skin are slightly weaker than the left side, indentation test (-), bilateral brachial plexus pull test (-), the skin feels no decline. Bilateral Hoffmann sign (-), pap sign (-), knee reflex (+ +), Achilles tendon reflex (+ +), triceps reflection (+ +), biceps reflex (+). Cervical vertebra activity is limited, cervical vertebra activity: left-handed 30°<=>40° right-handed; forward flexion 20°<=>20° posterior extension; left lateral flexion 20°<=>20° right flexion. Bilateral ankle clonus test (-), patellar clonus (-). Bilateral deltoid and biceps and triceps muscle IV level; Both hands hold the IV level, and the wrist side is extended to the IV level. Bilateral quadriceps muscle IV - and iliopsoas muscle IV - , pretibial muscle IV, leg triceps muscle IV, IV muscle power, length of the fibula. On the right side of the quadriceps muscle IV - and iliopsoas muscle IV - , pretibial muscle IV, leg triceps muscle IV, The length of the fibula muscle IV. Cervical MRI showed that the axial dentate process moved backward, the medulla oblongata and cervical curvature increased, and the corresponding signal in the medulla was less homogeneous.


Surgical name: Skull traction reduction + posterior cervical spinal decompression + bone graft + occipitocervical fusion.

Surgical step: After the patient’s anesthesia is successfully performed, the supine position is taken, the neck is in the overextended position, and the patient’s odontoid reduction is satisfactory. So, I give up the anterior operation. Take the prone position, bend the head and neck, and place the head on the head frame. The field is routinely iodophor disinfection, toweling, and skin care film. Surgical incision along the median longitudinal line of the neck, from the occipital trochanter to the C5 spinous process, subcutaneous and term ligaments, automatic retraction to retract the skin, subperiosteal dissection along the spinous process lamina, see C1 posterior arch loss, fusion with the occipital bone. C2, C3, C4 lamina and articular processes were revealed, see C2 and 3 conjoined vertebrae. The appropriate length of the occipital cervical fusion titanium plate is selected. After prebending, the patient’s skull is put in place. C2 and C4 bilateral pedicle holes were performed. After sounding, the screws are screwed in. Intraoperative fixation is stable. Then, the posterior margin of the occipital foramen is removed and the decompression is removed. After the autogenous iliac bone was trimmed, the wire was tied to the occipital bone and the C2 and 3 spinous processes. The broken bone pieces are implanted around. Flush and stop bleeding and check the gauze of the instrument. Place a rubber drainage tube and close the surgical incision in layers.

Figure 1: Preoperative X-ray showed atlanto-occipital osteosis and C2-3 fusion: a. anteroposterior view; b. lateral view; c. In MRI sagittal position, C2-3 vertebral body fusion, the occipital foramen is relatively narrow, the cervical spinal cord is compressed on the ventral side of the occipital neck, and the medullary cervical medulla becomes smaller.

**Figure 2:** Preoperative CT 3d reconstruction showed cranial depression, atlantoaxial occipital osteosis, increased atlantoaxial space, and atlantoaxial dislocation: a. coronal view; b. sagittal view; c. Cross sectional view.

**Figure 3:** Preoperative 3D printing model: a. rear view; b. front view; c. vertical view.


**Figure 5:** The postoperative X-ray showed that the internal fixation is in good position, the curvature of the craniocervical region is restored, and the atlantoaxial vertebra is restored well. a. Anteroposterior view of X-ray; b. Lateral view of X-ray; c. Postoperative chest wearing neck brace fixed for 3 months.

**Figure 6:** Postoperative CT three-dimensional reconstruction showed the recovery of curvature and atlantoaxial articulation in the craniocervical region: a. coronal view; b. sagittal view show atlantoaxial restoration; c. Sagittal position showing pedicle screw orientation; d. 3d reconstruction of the front view; e. 3d reconstruction of the rear view; f. 3d reconstruction of the left view.
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**Figure 7:** Postoperative 3D printing model: a. rear view; b. front view; c. vertical view.

### Supplement Table 1: The postoperative improvement rate based on ASIA sensory score.

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### Supplement Table 2: The postoperative improvement rate based on ASIA exercise score.

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Bibliography

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