Adjacent Segment Disease in a Patient With Klippel-Feil Syndrome and Radiculopathy: Surgical Treatment With Two-Level Disc Replacement

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ABSTRACT

Klippel-Feil syndrome (KFS) is a complex congenital condition characterized by improper segmentation of cervical motion segments that could contribute to undesirable adjacent segment degeneration. KFS patients have a strong tendency to present with disease in the adjacent segments. When this condition is present, anterior decompression followed by total disc replacement can be performed safely and can lead to good clinical results. This treatment has theoretical advantages compared with anterior decompression and fusion. Comparative studies and long-term follow-up are needed. Complications associated with fusion include loss of a motion segment, disc height loss, subsidence of the graft, progressive degenerative changes at the adjacent level, graft-related complications, and graft-site complications. Such new technologies as motion preservation spine arthroplasty represent attempts to avoid these complications.

Here we present a case report of a 62-year-old female patient with type I congenital fusion at the C5–6 level, with a history of neck pain and right radiculopathy at C5–7. X-rays and MRI show evidence of adjacent segment degeneration at levels above and below congenital fusion. The patient's preoperative visual analog score (VAS) for neck pain was 7 out of a possible 10, her score for right upper extremity pain was 8 out of 10, and her Neck Disability Index (NDI) was 32%. Surgical treatment consisted of anterior decompression and total disc replacement at both levels. At 1-year follow-up, the patient's VAS for neck pain was 2 out of 10, her VAS score for right upper extremity pain was 1 of 10, and her NDI was 9%.

Key Words Klippel-Feil syndrome, adjacent segment degeneration, cervical total disc replacement, motion preservation.
L2–3 between C5 and T1 in the right upper extremity. The symptoms were aggravated by extension and right side lateral rotation of the head. Anterior and lateral x-rays showed fusion type I of C5–6 levels, loss of cervical lordosis, and decreased height in C4–5 and C6–7 levels (Figure 1). Flexion–extension x-rays showed translational instability in the C4–5 level. MRI findings showed spinal stenosis at C4–5 and C6–7 levels, compression of the dural sac in both levels, and loss of normal disc height and hydration (Figures 2–4).

We treated the patient at our center using the anterior approach with decompression at C4–5 and C6–7 levels and application of total disc replacement M6 (Spinal Kinetics Inc., Sunnyvale, California, USA) in these two levels (Figure 5).

Postoperative follow-up at 1 year showed improvement in neck and right arm pain. The patient’s postoperative VAS at that time was 2 out of 10 for neck pain and 1 out of 10 for upper extremity pain. Her muscle strength also improved. She scored 5 out of a possible 5 in all muscular groups, and her NDI at 1-year follow-up was 9%. The patient was satisfied with these results. One-year postoperative x-rays showed recovery of cervical lordosis and return of normal movement of the motion segments treated with the disc arthroplasty (Figures 6 and 7).

DISCUSSION
The designation of Klippel-Feil syndrome includes a heterogeneous group of patients unified only by the presence of congenital synostosis of some or all cervical vertebrae. The condition occurs in approximately one in 40,000 to 42,000 births, with a slight female predominance of 3:2. The syndrome usually is diagnosed at a later age (>40 years of age) when neurologic, myelopathic, and biomechanical problems are present. Patients with progressive symptomatic segmental instability or neurologic compromise are candidates for surgical stabilization of the abnormal region of the cervical spine. Normally, this is obtained by anterior decompression and cervical fusion with very good documented results. However, there are two special concerns in this type of patient: adjacent segment disease in nonfused levels and loss of cervical motion. Robbins and Hilibrand10,11 chronicled 374 patients with a total of 409 anterior cervical arthrodoses. They found symptomatic
adjacent segment disease at a relatively constant incidence of 2.9% per year (0.0% to 4.8%).\textsuperscript{10,11} New disease was defined as new onset of myelopathy or radiculopathy significant enough to require surgery. Survivorship analysis projected that 25.6% of the patients (95% CI, 20% to 32%) who had an anterior cervical arthrodesis would have new disease at an adjacent level within 10 years after the operation. The most likely levels to develop adjacent segment disease were C5–C6 and C6–C7. Baba et al.\textsuperscript{12} assessed over 100 patients undergoing anterior cervical fusion for cervical myelopathy with an average of 8.5 years of follow-up. These authors observed that 25% of these patients subsequently developed new spinal canal stenosis above the previously fused segments. Gore and Sepic\textsuperscript{13} observed new spondylosis in 25% of 121 patients and progression of preexisting spondylosis in another 25% of patients who had previously undergone anterior cervical fusion with an average follow-up of 5 years.

Such motion preservation techniques as disc arthroplasty allow restoration of intervertebral disc height and neuroforaminal height and motion after anterior cervical decompression. These...
results seem to offer a better rationale for treatment in these patients. It is still possible that restoration of normal motion to an already diseased spinal segment in the cervical spine with cervical disc replacement will alter the natural rate of adjacent segment degeneration and, in fact, lessen it.

To the best of our knowledge only two studies have reported the use of total disc replacement in patients with KFS. McAfee\textsuperscript{14} reported on 33 patients who underwent a total of 32 porous coated motion (PCM) cervical arthroplasty procedures from C3–4 to C6–7: 15 were in a single level, 7 were in double levels, and 1 was in three levels. Three of these patients had KFS and had been treated with a variety of PCM with supplemental screw fixation in both components. At 9-month follow-up all patients were neurologically intact. A total of 79.9% of these cases had 15 percentage or more of improvement over preoperative scores on the Oswestry NDI, and 82.6% had greater than 20% improvement on VAS.\textsuperscript{14}

Pimenta et al.\textsuperscript{15} reported on 53 patients treated with PCM prosthesis. Among these patients, symptoms in two resulted from congenital fusion related to KFS. The average preoperative VAS was 85 points, NDI was 45%, and the average treatment intensity gradient test (TIGT) score was 11.6 points. At 1-year follow-up the mean VAS score was 20 points, the mean NDI was 15%, and the mean TIGT was 3.5 points. According to Odom criteria 57% reported excellent results.\textsuperscript{15}

Each of these studies includes a special report of KFS-related levels treated and the type of congenital fusion performed. We report here on a type I\textsuperscript{16} congenital fusion of the cervical spine with adjacent segment disease in the levels above and below in a patient who was successfully treated with cervical arthroplasty. One year after surgery, the patient’s x-rays showed excellent movement of the segments that were treated. One hopes that this type of treatment will decrease the rate of adjacent segment and improve cervical range of motion and patient satisfaction.

We have demonstrated in this case report that this technique is safe and successful in this type of patient. Complications related to the use of cervical disc replacement remain a concern. These complications include heterotopic ossification, persistent pain, prosthetic migration, segmental kyphosis, and device failure.\textsuperscript{17} Long-term follow-up and comparative studies with anterior cervical decompression and fusion are necessary to establish treatment guidelines.

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REFERENCES


