



Case Report

Two-level cervical disc arthroplasty in patients with Klippel-Feil syndrome: A case report and review of the literature

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ABSTRACT

Background: Klippel-Feil syndrome (KFS) is defined by multiple abnormal segments of the cervical spine with congenital synostosis of two or more cervical vertebrae. KFS patients who demonstrate progressive symptomatic instability and/or neurologic sequelae are traditionally managed with operative decompression and arthrodesis.

Case Description: A 44-year-old female with chronic neck pain and radiculopathy and a C7-T1 KFS presented with adjacent segment degenerative disc disease at the C5-6 and C6-7 levels. She was successfully managed with a two-level cervical disc arthroplasty (CDA).

Conclusion: Patients with KFS and disease at two contiguous, adjacent levels (e.g., cervical disc disease) may be safely and effectively managed with two-level CDA.

Keywords: Adjacent segment disease, Artificial disc replacement, Cervical disc arthroplasty, Congenital cervical fusion, Klippel-Feil syndrome, Motion preservation surgery

INTRODUCTION

The United States Food and Drug Administration investigational device exemption trial documented that 10 years following two adjacent level cervical disc arthroplasty (CDA), patients continued to show clinical improvement and preservation of motion at the operated levels.^[1]

In this case study, a 44-year-old female with a history of a C7-T1 Klippel-Feil syndrome (KFS) and two-level adjacent disc disease, successfully and safely underwent a contiguous two-level CDA (i.e., at the C5-C6 and C6-C7 levels).

CASE REPORT

History and examination

A 44-year-old female with a history of C7-T1 KFS presented with the chief complaint of neck and right-arm pain with radiculopathy. On examination, she had limited cervical flexion and extension, right paraspinal cervical tenderness, and weakness in the right C6 and C7 root distributions (both 4/5).

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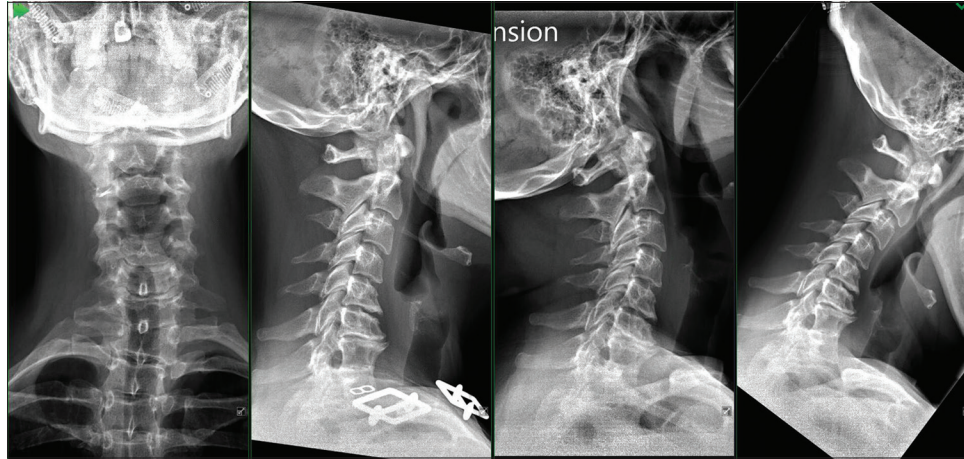


Figure 1: X-ray cervical spine with flexion and extension views revealing of advanced cervical degenerative disc disease at C5-6 and C6-7 with congenital fusion of C7-T1.

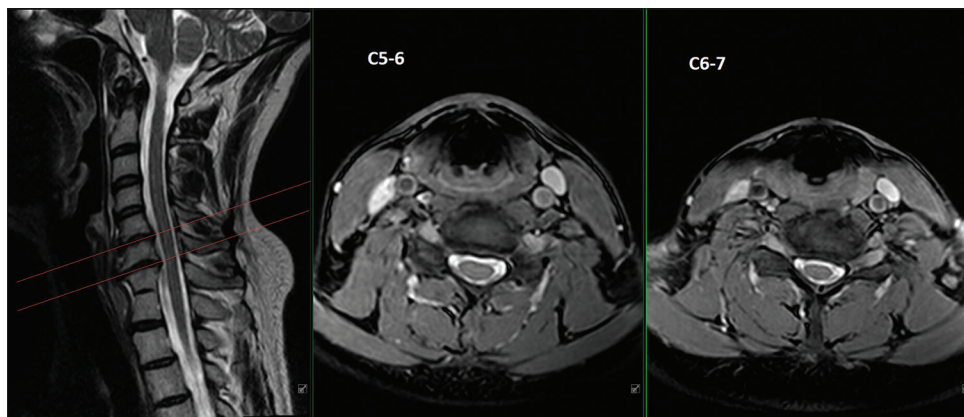


Figure 2: MRI cervical spine revealing of advanced cervical degenerative disc disease at C5-6 and C6-7 with congenital fusion of C7-T1.

Imaging

Cervical dynamic X-rays showed a loss of the normal cervical lordosis with a congenital KFS fusion at the C7-T1 level; dynamic studies confirmed no instability at that level [Figure 1]. In addition, the MR revealed bilateral, right greater than left, neuroforaminal stenosis at the C5-C6 and C6-C7 levels, without accompanying significant central stenosis [Figure 2]. The CT also confirmed the presence of the KFS congenital fusion at the C7-T1 level [Figure 3].

CASE DESCRIPTION

The patient underwent a two-level C5-C6 and C6-C7 CDA. A routine exposure was performed. Complete C5-C6 and C6-C7 discectomies, including resection of the posterior longitudinal ligament, were performed. In addition, the endplates at both levels were contoured and keels were cut to allow for implantation of the CDA devices [Table 1 and Figure 4].

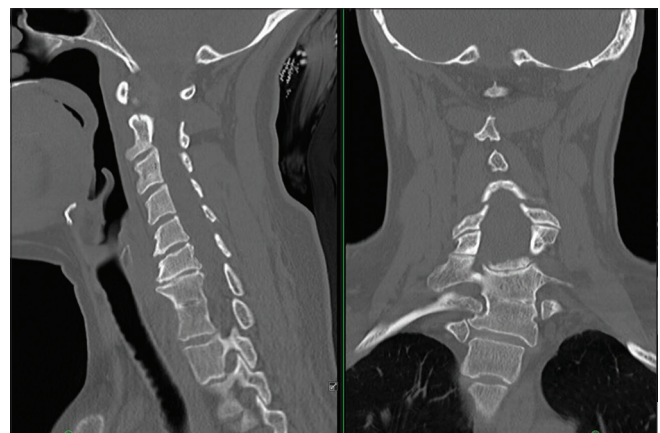


Figure 3: CT cervical spine revealing of congenital fusion of C7-T1 vertebral bodies.

Postoperatively, the patient regained full 5/5 strength of her right C6 and C7 root distributions. Formal upright vertical radiographs the next day, before discharge, confirmed adequate CDA positioning [Figure 5]. The patient returned

to work with restrictions and activity modifications 2 weeks later. At 8 postoperative months, her neurologic examination

was normal, and radiographically, the CDA devices were adequately positioned [Figure 6].

Table 1: Cervical disc arthroplasty implant sizes.

CDA implant	Height (mm)	Depth (mm)
C5-6	5	16
C6-7	5	18

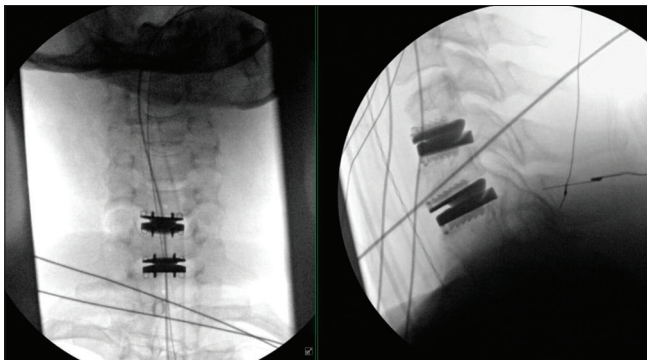


Figure 4: Final AP and lateral intraoperative fluoroscopic views confirming excellent placement of the cervical disc arthroplasty devices.

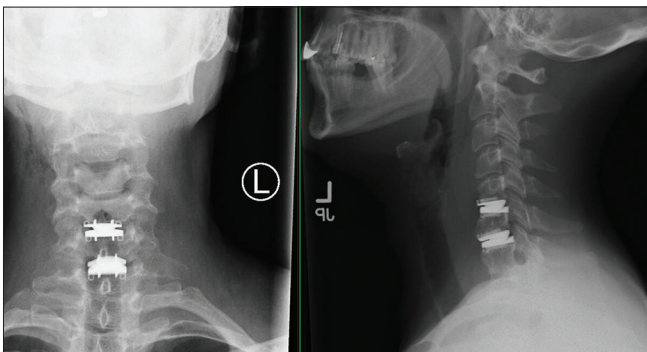


Figure 5: POD#1 AP and lateral cervical radiographs confirming excellent placement of the cervical disc arthroplasty devices.

DISCUSSION

Klippel-Feil patients with progressive symptomatic instability and/or neurologic sequelae are traditionally managed with ACDF. However, adjacent segment disease with the added loss of cervical range of motion is of particular concern in this patient population.

Symptomatic adjacent segment disease

Hilibrand and Robbins established that symptomatic adjacent segment disease occurs in 2.9% of patients/year and that 25% develop adjacent segment disease requiring additional surgery within 10 years following an ACDF.^[2] Therefore, motion-preserving CDA offers an alternative treatment modality.

Theory behind CDA

The theory behind utilizing CDA, specifically adjacent to a KFS, is to decrease the incidence of future adjacent cervical disc disease. Gornet and Lanman's multicenter data demonstrated the superiority of two-level CDA over ACDF at 10 postoperative years; the overall success was 80.4% for CDA versus 62.2% for ACDF.^[1] Postoperatively, the patient's radiographs showed excellent movement of the segments treated.

A recent meta-analysis of 19 trials found that CDA was superior to ACDF in terms of overall neck disability index (NDI), neurological recovery of function, higher 36-Item Short Form Health Survey (SF-36) results, higher patient satisfaction, greater range of motion, and fewer secondary operations ($P < 0.05$).^[3] Another meta-analysis involving 650 patients, observed that CDA was superior to ACDF for two

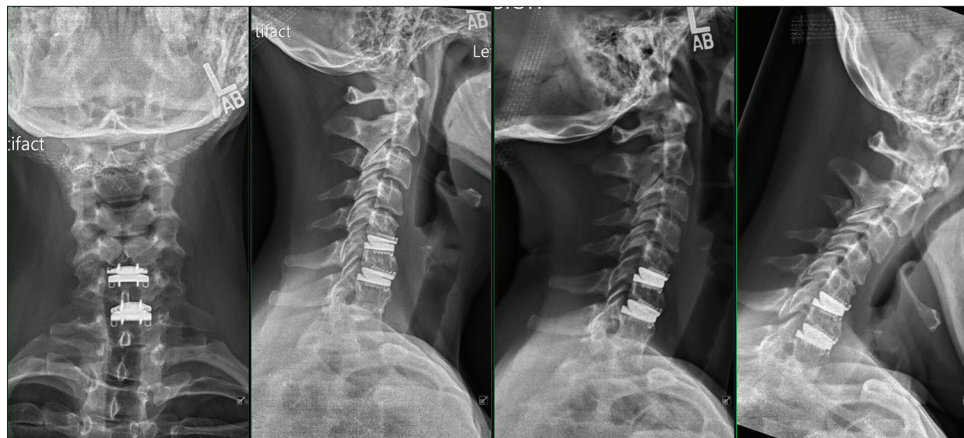


Figure 6: One-month postoperative AP and lateral cervical radiographs confirming excellent placement of the cervical disc arthroplasty devices and motion through flexion and extension.

Table 2: Meta-analysis comparing CDA versus ACDF for two contiguous level cervical degenerative disc diseases.

Mean blood loss		CDA			ACDF			Weight	Std. mean difference IV, fixed, 95% CI
Study or subgroup	Mean	SD	Total	Mean	SD	Total			
Cheng, 2011	100.2	29.4	14	150.2	41.2	17	21.9%	-1.34 (-2.13, -0.55)	
Hou, 2013	168	51	31	210	61	88	78.1%	-0.71 (-1.13, -0.29)	
Total (95% CI) $P < 0.00001$			45			105	100.0%	-0.85 (1.22, -0.48)	
Neck disability index		CDA			ACDF			Weight	Std. mean difference IV, fixed, 95% CI
Study or subgroup	Mean	SD	Total	Mean	SD	Total			
Cheng, 2011	50.6	6	14	50.1	5.8	17	7.3%	0.08 (-0.63, 0.79)	
Davis, 2015	36.5	21.3	225	28.5	18.3	105	61.0%	0.39 (0.16, 0.62)	
Hou, 2013	19.5	12.	31	18.7	12.3	88	21.8%	0.06 (-0.35, 0.47)	
Jawahar, 2010	44.9	2.6	16	43	2.9	6	3.9%	0.68 (-0.28, 1.65)	
Kim, 2009	8	0.9	12	7.8	1.3	28		Not estimable	
Total (95% CI) $P = 0.002$			286			216	100.0%	0.31 (0.12, 0.50)	
Reoperation		CDA			ACDF			Weight	Risk ratio M-H, fixed, 95% CI
Study or subgroup	Events	Total	Events	Total	Events	Total			
Cheng, 2011	0	14	0	17				Not estimable	
Davis, 2015	9	225	16	105	93.0%			0.26 (0.12, 0.57)	
Grob, 2009	0	19	4	89	7.0%			0.50 (0.03, 8.92)	
Total (95% CI) $P = 0.0009$		258		211	100.0%			0.28 (0.13, 0.59)	
Total events	9		20						
Adjacent segment disease		CDA			ACDF			Weight	Risk ratio M-H, fixed, 95% CI
Study or subgroup	Events	Total	Events	Total	Events	Total			
Davis, 2015	93	225	90	105	80.2%			0.48 (0.41, 0.57)	
Hou, 2013	5	31	37	88	12.6%			0.38 (0.17, 0.89)	
Jawahar, 2010	3	16	1	6	1.0%			1.13 (0.14, 8.82)	
Kim, 2009	4	12	16	28	6.3%			0.58 (0.25, 1.38)	
Total (95% CI) $P = 0.00001$		284		227	100.0%			0.48 (0.4, 0.58)	
Total events	105		144						

contiguous level diseases with regard to reduced blood loss, fewer reoperations, less adjacent segment disease, and better NDI [Table 2].^[6] Furthermore, when Wu *et al.*^[5] compared the results of CDA versus ACDF (i.e., in five studies) adjacent to a level of previous fusion, they found that CDA had improved clinical outcomes, better preservation of segmental motion, and comparable complication rates. McAfee cited similar findings.^[4]

CONCLUSION

Here, we have demonstrated that a patient with a C7-T1 KFS and two-level adjacent segment disease (e.g., C5-C6 and C6-C7) could be safely and successfully managed with a contiguous, two-level C5-C6/C6-C7 CDA.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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