Adjacent Segment Level Disease in Klippel-Feil Syndrome Patients with Congenital Cervical Fusion in the Setting of Anterolisthesis: Should Management be Altered in the Presence of Anterolisthesis?
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Introduction
The goal of our study was to determine the incidence of adjacent segment level pathology preoperatively in patients with Klippel-Feil syndrome (KFS) and compare it to the incidence in patients who have undergone anterior cervical discectomy and fusion. Secondly, we hypothesized that patients with KFS and anterolisthesis would be more likely to develop symptomatic and radiological adjacent segment level disease.

Methods
Twenty patients with Klippel-Feil syndrome from a single institution were followed for 33 months; range 4e108 months. Serial imaging and follow-up (serial visits) were used to determine adjacent segment level disease. Patients were separated in two categories: those with anterolisthesis adjacent to their congenitally fused segments and those without.

Results
Twenty patients with an average age of 9.5 yrs were identified. The average follow-up period was 33 months; range 4e108 months. Nineteen patients received a diagnosis of KFS during incidental imaging. One patient was diagnosed as part of a workup for myelopathy caused by a disc herniation adjacent to two fused segments. Eight patients had no listhesis in cervical levels adjacent to fused vertebral segments while 12 had listhesis. The two most common levels of listhesis were at C3eC4 and C4eC5 (five patients each). Five patients in the listhesis group had an increase in listhesis through the follow-up period. There were no differences in the development of symptoms between patients who had and did not have listhesis. All nonoperative patients were asymptomatic at last follow-up.

Learning Objectives
- The incidence of adjacent segment level pathology preoperatively in patients with Klippel-Feil syndrome (KFS) differs from patients who have undergone surgical fusion in the cervical spine
- Our follow-up was limited, but this study does not support the assumption that a congenitally fused segment predisposes a patient to a rate of adjacent segment level disease similar to patient who has undergone an anterior cervical discectomy and fusion. We found no increased risk of symptomatic adjacent segment level disease in KFS patients who had anterolisthesis adjacent to a congenitally fused segment.

References