Massive Diminution of the Cervical Spine: One Case Report

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Key words: Klippel-Feil anomaly, neck, anatomy

SUMMARY

The authors report a 13-year-old female with shunted hydrocephalus who when evaluated for neck pain, was found to have not only fusion of cervical vertebrae but significant diminution of the cervical spine. Following our review of the classification of KFA and our case report, the current interpretation of Klippel and Feil's original description of KFA should be re-evaluated, and/or a new class of KFA used, that of complete or partial cervical vertebrae agenesis and associated abnormalities, should be considered in the classification of this anomaly.

INTRODUCTION

In 1912, Maurice Klippel and Andre Feil were the first to describe congenital fusion of the cervical spine [Klippel-Feil anomaly (KFA)] (Klippel and Feil, 1975). KFA has an incidence of up to 0.5% of live births (Brown et al., 1964; Gray et al., 1964; Le Double, 1912; Luftman and Weintrub, 1953). Gjorup and Gjorup (1964) reviewed all of the radiographic cervical spine films in a single hospital in Copenhagen, and determined an incidence of 0.2 cases per 1000 people. This anomaly has been characterized in patients by a triad of symptoms, involving a short neck, decreased range of motion (ROM) in the cervical spine, and a low hairline. Fewer than 50% of patients have all three elements of this triad therefore, in its current usage, the term KFA refers to persons with congenital fusion of the cervical vertebrae (Raas-Rothschild et al., 1988). KFA is most apparent after normal spinal ossification nears completion in the young child. Patients presenting with upper cervical spine involvement tend to present clinically at an earlier age than those whose involvement is lower in the cervical spine, but regardless, decreased range of motion of the cervical spine is the most frequent clinical finding.