**Klippel-Feil syndrome**

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**Introduction**

Klippel-Feil syndrome is a rare congenital disease initially reported by Maurice Klippel and Andre Feil from France in 1912 [1]. It is characterized by the congenital fusion of at least two of the seven vertebrae of cervical spine [2]. It is believed to result from the faulty segmentation along the embryo's developing axis during 3-8 weeks of gestation. The classic triad consists of short neck, low hair line and restricted neck movements, presenting in less than 50% of patients with this syndrome. It is detected throughout the life, often as an incidental finding.

**Case report**

This is a plain cervical spine X-ray (Figure 1) of a 33-year-old male, with no significant past medical history, who came with complaints of right shoulder and left forearm pain with some neck pain after he was involved in a motor vehicle accident. Examination revealed tenderness of right shoulder joint with some restriction of movements, left mid forearm tenderness and mild posterior midline neck tenderness. There were no other injuries. Detailed systemic review including central nervous examination was normal. X-rays were done for right shoulder joint, left forearm and cervical spine. Shoulder and forearm X-rays were normal. X-ray of cervical spine showed fusion of C4-5 spinal bodies along with posterior neural arches. Diagnosis of Klippel-Feil syndrome was made. He was discharged with advice on avoiding neck injuries, and was suggested to report back immediately in case of any abnormal sensations or lack of sensations in his upper limbs.

**Discussion**

Andre Feil proposed a classification scheme for Klippel-Feil syndrome in 1919 [3]. The classification is based on cervical, thoracic and lumbar spine malformations as:

- **Type I**: a massive fusion of cervical spine;
- **Type II**: the fusion of 1 or 2 vertebrae;
- **Type III**: the presence of thoracic and lumbar spine anomalies in association with Type I or Type II Klippel-Feil syndrome.

Samartzis and his colleagues suggested a new classification system in 2006 that specifically addressed cervical spine anomalies as [4]:

- **Type I**: single congenitally fused cervical segment;
- **Type II**: multiple noncontiguous, congenitally fused cervical segments;
- **Type III**: congenital fusion of more than two cervical vertebrae.

**Keywords**

Klippel-Feil syndrome; Cervical vertebrae; X-rays; Case reports.

【关键词】克利佩尔-费尔综合征; 颈椎; X线; 病例报告
Type III: Multiple contiguous, congenitally fused cervical segments.

The incidence is estimated at about one in 42,000 births ($^5$). Some have reported equal incidence in both sexes ($^4$) where as others have reported unequal incidence in both sexes ($^7$).

Associated abnormalities include scoliosis, spina bifida, cleft palate, respiratory problems, heart malformations, anomalies of kidney and ribs. The prognosis is good in the absence of associated abnormalities.

Persons with Klippel-Feil syndrome and cervical stenosis should be made aware of the potential for sustaining a neurologic deficit after minor trauma. Activities that can injure the neck should be avoided.

Klippel-Feil syndrome is treated symptomatically, but surgical treatment may be required for patients not responding to symptomatic treatment. Cervical disc arthroplasty using Bryan cervical disc prosthesis (Figure 2) may be required ($^8$). Surgical options are adopted to maintain the range of movement of two adjacent fragments ($^9$).

**Conclusion**

Most of the time Klippel-Feil syndrome is an incidental finding treated symptomatically and patients can be discharged with precautionary advice related to neck movements, but few cases may require surgical correction.

**Disclosure**

No authors report any conflict of interest.

**References**


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