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# Challenges for subarachnoid block in a patient with klippel feil syndrome: A case report

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**Abstract**
Klippel Feil syndrome is one of the rarest congenital anomalies with newborns worldwide, with a slight female predominance. Characterised by abnormal fusion of two or more cervical vertebrae. Klippel-Feil syndrome presents with the classic triad of a short neck, low posterior hairline, and limited neck mobility. It presents a unique perioperative challenge to the anaesthesiologist as both the airway management and central neuraxial blockade are anticipated to be difficult due to fusion of the vertebrae and the presence of other associated congenital anomalies.
27 year old male with Klippel Feil syndrome, weighing 55 kg with a height of 154.94 cm and BMI of 22.9 kg/m2 presented in our emergency with left irreducible indirect inguinal hernia and was planned for hernioplasty. He had been diagnosed previously as Type III Klippel Feil syndrome having the classical triad, which is quite rare. He was planned under subarachnoid block, which was quite challenging. Surgery was uneventful and the patient made an excellent recovery.
In individuals with Klippel-Feil syndrome, administration of subarachnoid blockade necessitates meticulous preoperative orchestration, given the intricate anatomical aberrations. Vigilant preparedness for potential airway and neuraxial complexities is imperative to facilitate safe and efficacious anaesthetic intervention.

**Keywords:** Klipple feil syndrome, subarachnoid block, triad, sprengel deformity

## Introduction
Klippel Feil syndrome is one of the rarest congenital anomalies with newborns worldwide, with a slight female predominance of 1 in 42,000 newborns worldwide [1]. It is characterised by abnormal fusion of two or more cervical vertebrae [2]. Klippel-Feil syndrome presents with the classic triad of a short neck, low posterior hairline, and limited neck mobility [2]. It presents a unique perioperative challenge to the anaesthesiologist as both the airway management and central neuraxial blockade are anticipated to be difficult due to fusion of the vertebrae and the presence of other associated congenital anomalies.

## Case report
27-year-old male with a known case of Klipple Feil syndrome diagnosed one year back (weight of 55 kg, height: 154.94 cm and body mass index of 22.9 kg/m²) non-smoker, non-alcoholic presented with chief complaint of abdominal pain for 3 days and left irreducible inguinal swelling since 2 days. He also had congenital glaucoma in left eye and scoliosis with decreased intervertebral spaces and Sprengel's scapula deformity (25-35%) [3]. There were no other associated congenital anomalies and he had no functional limitations with METS>4.
On pre operative examination-

- Hemodynamically stable vitals
- Normal general physical examination
- Congenital glaucoma in left eye
- Systemic examination - normal

### Airway examination

- Modified Mallampati grade III
- Mouth opening- 2 finger
- Thyromental distance- 2 finger
- Neck extension- limited (20 degrees)

- Upper lip bite test - class III
- Spine examination- scoliosis with decreased intervertebral spaces
- Left sprengel's scapular deformity

**Investigations**

- Blood investigations- normal
- Cervical vertebrae showed decreased intervertebral space with fusion of C4-C5 vertebrae
- Lumbar vertebrae shower scoliosis with decreased intervertebral spaces

**Anaesthetic management**

- Patient and his family was counselled and explained about the procedure, its complications and difficulties related to KFS and a written informed consent was taken.
- Hernioplasty was planned under subarachnoid block, however complete arrangement for difficult airway like airway emergency cart, fibro optic bronchoscope were also made ready.
- With patient in the sitting position, subarachnoid block was initially attempted through midline approach by resident, first in L4-L5 and then L3-L4 interspaces using 25G Quincke needle and was unsuccessful.
- Subsequent attempt was made by senior anaesthesiologist via paramedian approach in L3-L4 interspace and was successful.
- After five attempts subarachnoid block was then achieved with a total of 3ml (-2.8 mL of 0.5% hyperbaric bupivacaine with 10 mcg fentanyl).
- The patient developed sensory block of T4 dermatome in 2 minutes, after which his head was elevated using pillow.
- Intraoperative - vitals were stable with one dose of hypotension for which injection mephentermine 6 mg bolus was administered intravenously.
- Total fluid administered was 1200 ml of balanced salt solution
- Total duration of surgery was 2 hours
- At the end of surgery the block levelmwas motor -M2 and sensory -T10
- Surgery was uneventful and the patient made an excellent recovery.

**Discussion**

KPS is inherited autosomal dominant disorder characterised by a triad of short neck, severe restriction of neck movements and a low posterior hairline. It is commonly associated with various other congenital anomalies: scoliosis, sprengel deformity, cataract, renal abnormalities, deafness, synkinesis, and congenital heart diseases [4]. This case highlights the anaesthetic challenges associated with Klippel-Feil syndrome, particularly the technical difficulty of neuraxial blockade due to vertebral fusion and scoliosis. In this patient, successful subarachnoid block required multiple attempts using a paramedian approach. Thorough preoperative assessment was essential, given the potential for airway difficulties and other associated congenital anomalies.

**Anaesthetic considerations**

- Positioning difficulties may arise due to fusion of cervical, thoracic, or lumbar vertebrae, often accompanied by scoliosis.
- Exaggerated lumbar lordosis can make achieving optimal positioning for neuraxial blocks more challenging.
- Fused spinal segments increase the risk of transient neurological deficits, even after minor trauma.
- Spinal anomalies such as kyphosis and abnormal lumbar curvature can lead to unpredictable intrathecal drug spread.
- Fused thoracic and lumbar vertebrae can make administration of neuraxial anesthesia technically difficult

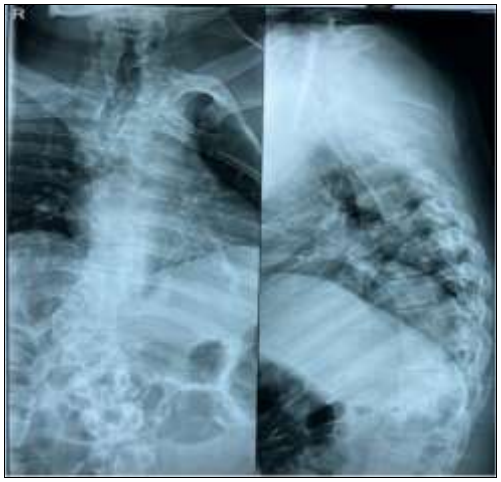

**Fig 1:** Lumbar scoliosis

**Fig 2:** Sprengels scapular deformity

**Fig 3:** Thyromental Distance 2 Fingers

**Fig 4:** Short Neck And Limited Neck Mobility

**Fig 5:** decreased intervertebral space with C4-C5 Fusion

## Conclusion

Neuraxial anaesthesia can be safely performed in cases of KFS undergoing lower limb surgeries or abdominal surgeries where low level block is required. In individuals with Klippel-Feil syndrome, administration of subarachnoid blockade necessitates meticulous preoperative evaluation, in view of anatomical difficulties. Vigilant preparedness for potential airway and neuraxial complexities is essential to facilitate safe and efficacious anaesthetic intervention. However there is need for careful preoperative clinical and radiological spine evaluation to identify patients with potential difficulty and planning to manage the anticipated problems in performing subarachnoid block.

## Declaration of Patient Consent

The author confirms that informed consent was obtained from the patient for publication of clinical details and images. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

## Conflicts of Interest

The authors declare that they have no conflicts of interest.

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