Abstract: Klippel-Feil syndrome is an inherited autosomal dominant condition and is associated with multiple congenital anomalies including short neck, complete fusion of the cervical spine and severe limitation of neck movements which can cause difficult airway. The deformity of spine pose a challenge for positioning and regional anaesthesia. In this article, we present a case of a CTEV baby scheduled for an elective surgery successfully managed with intravenous sedation and caudal block.

Keyword: Klippel-Feil syndrome, CTEV, Difficult Airway.

Introduction
Klippel-Feil syndrome is a clinical triad consisting of fusion of cervical vertebrae, with associated shortening of the neck, restricted head motion, and a low posterior hairline. These patients have potential cervical instability and are at high risk for neurological damage during laryngoscopy, intubation and positioning, and thus pose a great challenge to the anesthetist. We report the anesthetic management of CTEV repair in a patient with Klippel-Feil syndrome.

Case Report
A 5 month old female baby weighing 4 kg was referred to our hospital for the repair of CTEV. In the preanesthetic evaluation, the child was found to have short neck, torticollis, posterior cleft palate, limitation of cervical extension and rotation. X-ray of cervical spine revealed fusion of C2-C3 vertebrae. Echocardiogram showed patent foramen ovale. CT brain showed flat and elongated pons. Fundus Examination showed deep and elongated cup. Ultrasound abdomen, ENT Examination and laboratorial test were normal.

Anesthesia for a child with klippel – feil syndrome: We planned intravenous sedation with caudal block, as it was lower limb surgery with anticipated difficult intubation. The baby was positioned supine on the table. Patient was connected to pulse oximeter, NIBP, ECG to be monitored continuously. An intravenous line was started on the dorsum of the right hand. The baby was premedicated with 0.1 mg of Atropine intravenously and anesthesia was induced with 50% N2O in oxygen with incremental dose of sevoflurane.
with maximum concentration of 5%. 0.25% Inj. Thiopentone sodium 10 mg was administered slowly via intravenous route. Trial laryngoscopy was done which showed Cormack Lehane Grade 2. Caudal block was performed with 4ml of 0.25% of Bupivacaine. Patient was positioned supine for the planned surgery. Spontaneous ventilation was maintained with 50% N2O in oxygen with 2% sevoflurane using mask connected to Jackson – Rees modification of Ayre’s T Piece. Duration of surgery was 40 min. Intraoperative blood loss was insignificant. After the procedure, the baby was given 100% oxygen through mask for 5 min. Recovery was good. The baby was subsequently shifted to the ICU for observation and had no adverse neurological sequelae.

Discussion:
There are 3 types of Klippel-Feil Syndrome. Type I is an extensive abnormality where elements of several cervical and upper thoracic vertebrae are incorporated into a single block. Type II is the most common variant and includes failure of complete segmentation occurring at one or two cervical interspaces. Type III variant includes Type I or II deformities with coexisting segmentation errors in the lower thoracic or lumbar spine. These babies may have associated other systemic anomalies of which the major concern for anaesthesiologists include skeletal system: Kyphoscoliosis, Sprengel deformity, Atlanto Occipital fusion which make airway difficult.
CardioVascular System: Ventricular septal defects, Patent ductus arteriosus, Mitral valve prolapse, Bicuspid aortic valve and Coarctation of the aorta which make preoperative cardiac evaluation mandatory. Other anomalies include Cleft palate, mandibular malformations and micrognathia which also complicate airway management. Hence, there is more chance of spinal cord trauma during laryngoscopy, intubation, and positioning of the patient. In these patients with anticipated difficult airway, Fiberoptic-guided intubation is the method of choice. However, the unavailability of fiberoptic equipment prompted us to perform trial laryngoscopy with inhalational induction. Sevoflurane is the best inhalation agent for induction as it is less irritant to the airway and also has low blood gas coefficient making induction faster. The use of LMA has been found to be a suitable alternative to endotracheal tube, but its use may be associated with more chance of airway obstruction and displacement. Careful preoperative evaluation to exclude associated congenital anomalies, avoidance of manipulation during laryngoscopy, intubation, and positioning, and better understanding of associated pathophysiology may improve the outcome in patients with Klippel-Feil syndrome, who require general anaesthesia. Actually 0.25% Inj. Thiopentone sodium 10 mg was given along with Sevoflurane 5% prior to laryngoscopy. It was wrongly printed in previous submitted paper. I deeply regret the inconvenience caused.
PATIENT AFTER CTEV REPAIR AND POP CAST

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