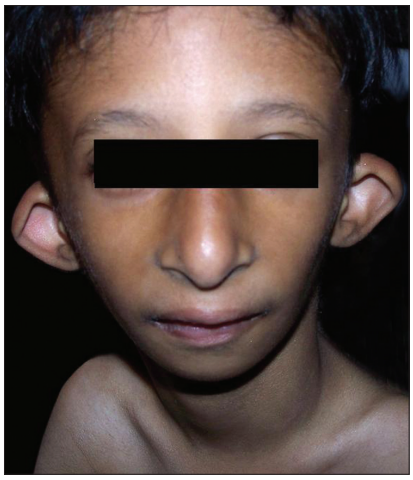

Figure 1: Facial profile of the patient

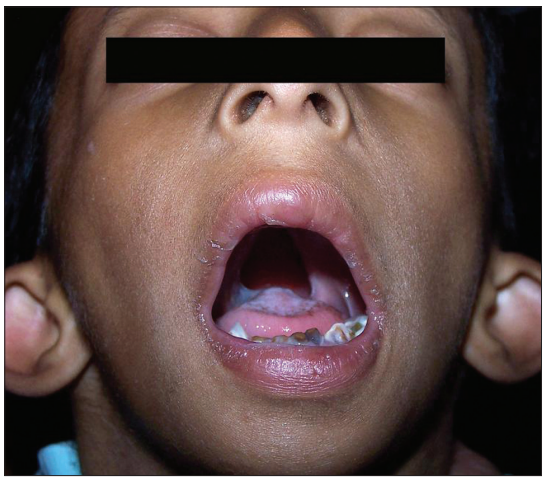

Figure 2: Cleft palate

Anesthetic management of a child with Seckel syndrome for multiple extractions and restoration of teeth

Sir,

Seckel syndrome is a rare autosomal recessive disorder. It is characterized by severe intrauterine and postnatal growth retardation, low birth weight, severe microcephaly, craniofacial dysmorphism with characteristic bird headed appearance, prominent beaked triangular nose, micrognathia and variable mental retardation.[1] Other multiple anomalies associated are cleft lip and palate, club foot, scoliosis, gastrointestinal malformations, multiple skeletal malformations, cardiovascular, endocrine, hematopoietic and central nervous systems abnormalities.[2-5]

There are several reported dental malformations in this syndrome for which patients may frequently require general anesthesia.[1,6] Although many cases of Seckel syndrome are described in literature only a few have been reported who required anesthesia.[7-11] We report the anesthetic management of a patient of Seckel syndrome scheduled for multiple extractions and restorations of teeth.

A 5½ kg, five-year-old boy, 75 cm in height, was scheduled for multiple extractions and restorations of teeth. He was born to healthy non-consanguineous parents at full term with birth weight of 900gm. At the age of three years he was diagnosed as a case of Seckel syndrome. The facial features were characteristic of the syndrome which included severe microcephaly, facial dysmorphism, beak like nose, prominent low set ears, micrognathia and cleft soft palate [Figures 1 and 2]. Mouth opening was adequate although the airway was modified Mallampati grade III. Patient also had spina bifida, syndactyly, undescended testis and growth retardation. Dental examination revealed dental hypoplasia and multiple carious teeth, requiring extractions and restorations. The cardiac and respiratory systems were within normal limits. The blood investigations (hematological and biochemical), chest radiograph and electrocardiogram (ECG) were normal.

Preoperative heart rate was 110/minute and blood pressure was 90/40 mm Hg. No premedication was administered to the child. Difficult airway cart was kept ready. After establishment of monitoring (ECG, peripheral oxygen saturation and noninvasive blood pressure), anesthesia was induced with inhalation of 5-8% sevoflurane in 100% oxygen using a size 0 Rendal Baker facemask and modified Jackson Rees circuit. Intravenous (IV) access was established with 24 G cannula at third attempt because of fragile veins. After

ensuring satisfactory mask ventilation, suxamethonium 1.5mg/kg was administered IV to facilitate endotracheal intubation. Laryngoscopy revealed Cormack Lehane grade III. Trachea was intubated with 4.0 mmID uncuffed tube using a stylet and fixed at 13 cm. Fentanyl 5μg was administered for analgesia. Anesthesia was maintained with O_2 (40%) N_2O, sevoflurane (with cumulative MAC between 1-1.2) and atracurium. IV paracetamol 10mg/kg was administered in the maintenance fluid. The procedure lasted for 2.5 hours and the intraoperative course was uneventful. After the procedure, muscle relaxant was reversed and the trachea was extubated. Postoperative course was uneventful and no episode of aponeic spells was observed in postoperative period.

There are multiple perioperative problems associated with Seckel syndrome. Anesthesia may be challenging primarily because of the difficulty in airway and venous access. A dysmorphic face with a receding chin and a relatively big nose could also render mask ventilation difficult.[11] Dental malocclusion, receding chin, high palatal arch could be the warning signs for the possibility of difficult intubation.[11] Although we were able to maintain mask ventilation without any difficulty we were prepared to handle the reported difficulty of mask fitting with an appropriate size laryngeal mask airway and the fiberoptic bronchoscope was kept ready in case intubation failed with direct laryngoscopy using stylet.

Age and weight of the child may fail to predict the tube size. The variation in tracheal tube size ranges up to 1-1.5 sizes smaller than the predicted size. . The tracheal tube size predicted by the chronological age in our patient was 5 mm ID [(Age in years + 16) divided by 4], but we could only intubate the trachea with 4.0 mm ID diameter tube which corresponded to that predicted by weight (five and half kg).

We used titrated doses of all the drugs. The anesthetic drugs used should be short acting. We used atracurium as muscle relaxant as prolonged neuromuscular recovery is reported with vecuronium in a child with this syndrome.[10] We did not use any premedication and used fentanyl for analgesia. There were no postoperative aponeic spells. Regional techniques and non-steroidal anti inflammatory drugs should be preferred for analgesia wherever possible.

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Quick Response Code:	Website: www.joacp.org
	DOI: 10.4103/0970-9185.98361