C-MAC® pediatric D-blade for intubation in a child with Hurler syndrome

Dear Editor,

Mucopolysaccharidoses (MPS) are a group of lysosomal storage disorders, associated with airway problems. An untreated case of Hurler syndrome (MPS-I) poses maximum difficulty during airway management. The incidence of difficult airway and failed intubations in MPS-I are 54% and 23%, respectively.[1] We describe here a successful use of the C-MAC® pediatric D-blade (Karl Storz, Tuttlingen-Germany) for intubation in a child with MPS-I.

A 6-year-old male child, weighing 13 kg, a diagnosed case of MPS-I, was scheduled for umbilical hernia repair. He had a history of noisy breathing, preference to sleep in the prone position and mental retardation. General examination of the child revealed short stature, cloudy cornea, coarse facial features, large tongue, modified Mallampati (MMP) grade-3, short neck and a large head [Figure 1]. Neck x-ray and echocardiography were normal.

In the operation room, difficult airway cart was kept ready and two senior anesthesiologists were present. Sedative premedications were avoided and glycopyrrolate 50 μg was given intravenously. Routine monitors (ECG, SPO_2 and Non-invasive blood pressure) were attached. The child was gradually induced with sevoflurane. Facemask ventilation was not adequate even with the two-hand technique and an oro-pharyngeal airway. So, a size 2 Ambu Laryngeal Mask was inserted for ventilation. After ensuring adequate ventilation through laryngeal mask, fentanyl 15 μg and Propofol 20 mg were given. Laryngoscopy with C-MAC® pediatric D-blade revealed a percentage of glottic opening (POGO) score of 90%. The child was intubated with a styletted 4.5 mm ID cuffed endotracheal tube (ETT) on the first attempt. For analgesia, a single dose lumbar epidural injection with 0.25% ropivacaine 6 mL and intravenous paracetamol 200 mg were used. The surgery was uneventful and the child was extubated when fully awake.

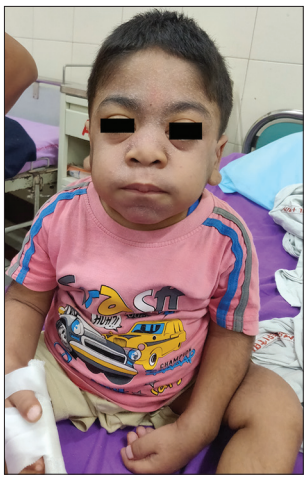

Figure 1: Child with Hurler syndrome

Hurler syndrome is associated with a very high incidence of failed intubations. Airway management has become better with the advent of enzyme replacement therapy, hematopoietic stem cell transfer, and newer intubation devices.[1] The recently introduced C-MAC® pediatric D-blade is one such device specially designed for syndromic children. D-blade has a curvature of 75° thereby increases the field of vision.[2] The hyper angulated blade is particularly useful in children with large tongue, higher MMP grades, and an anterior larynx. There is a high incidence of craniocervical junction stenosis and atlanto-axial instability in Hurler syndrome.[3] In such case, D-blade facilitates intubation with neck in the neutral position and minimal lifting force thereby preventing atlantoaxial dislocation.[2]

The common problem associated with hyper angulated blades is difficulty in negotiating the ETT in spite of a good glottic view. This can be avoided by shaping the styletted ETT similar to that of the D-blade curvature and inserting along its surface.[4] Even though we did not face any difficulty with the video laryngoscope, we were ready with fiberoptic guided intubation through laryngeal mask as a backup plan.[5] Ours is the first report on the pediatric D-blade being used for Hurler syndrome with difficult airway. We propose that C-MAC® pediatric D-blade is a useful aid in anesthesiologists' airway armamentarium while managing a syndromic child with difficult airway.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. 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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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