Literature research shows that infusion of noradrenalin and dobutamine before induction is effective in countering the depressive effect of anesthetic drugs. Further, cardio-stable and short-acting drugs such as etomidate, fentanyl, and vecuronium are employed as part of anesthetic regime. Noninvasive continuous cardiac output monitoring is used to evaluate the ventricular performance in response to fluid therapy, inotropes, and prone positioning. Prone position leads to increased intrathoracic pressure reducing venous return, and decrease in cardiac output (CO) and cardiac index (CI), which is exaggerated in DCM patients.

A noteworthy point with regard to pacing in these patients: preoperative conversion to asynchronous mode of CRT-P is necessary, in order to prevent pacemaker inhibition by electromechanical interference and use of bipolar cautery and also prevent pacemaker malfunction. For AICD, since these patients are not pacemaker-dependent, simple deactivation of the ICD intraoperatively may suffice. Additionally, AED pads should be placed before induction in the anterior–posterior configuration, so that the cardioversion/defibrillation current would not cross the pacemaker path.

**Conclusion**

Anesthetic management of patients with DCM on CRT device poses unique challenge for the anesthesiologist, with strong likelihood of catastrophic hemodynamic perturbations, dysrhythmias, and even sudden cardiac arrest. Particularly in surgery in prone position with limited access to the patient in the event of a catastrophe, meticulous planning, vigilant monitoring, adequate preparation with anterior–posterior configuration of transthoracic pads, judicious use of pharmacological agents, and tailor-made anesthetic regime, can lead to a favorable outcome.

**Conflict of Interest**

None declared.

**References**


**Case Report**

**Management of a Difficult Airway Scenario in a Case of Hurler’s Syndrome with a D-Blade Video Laryngoscope**

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Hurler’s syndrome is a rare genetic disease characterized by progressive multiorgan accumulation of glycosaminoglycans. It is associated with progressive craniofacial, skeletal, and cardiac involvement, which increases the risk of anesthesia. Patients with Hurler’s syndrome could present as the worst airway management problem an anesthesiologist could deal with due to abnormal upper airway anatomy and limited neck manipulation, owing to the atlantoaxial instability. We report a case of difficult airway scenario of a child with Hurler’s syndrome, leading to an apparent cannot ventilate cannot intubate scenario, which was managed successfully with the help of a C-MAC video laryngoscope with unique D-blade. In Hurler’s syndrome, C-MAC with D-blade is an excellent tool in establishing an airway in a pediatric difficult airway scenario. Moreover, D-blade C-MAC could be considered as the primary tool for establishing an airway in pediatric patients with Hurler’s syndrome.

Introduction
Hurler’s syndrome, a type 1 mucopolysaccharidosis (MPS 1H), is an uncommon genetic disease caused by deficiency of a specific lysosomal enzyme, a-1-iduronidase, which is required for a normal degradation of glycosaminoglycans and characterized by progressive multiorgan accumulation of the same.1 It is typically associated with progressive craniofacial, skeletal, and cardiac involvement, which increases the risk of anesthesia.1,2 We report a case of a difficult airway scenario of a child with Hurler’s syndrome, leading to an apparent cannot ventilate cannot intubate scenario, managed successfully with the help of a C-MAC video laryngoscope and pediatric D-blade. Informed consent was obtained from the parents prior to submission for publication.

Case Report
A 2-year-old girl, with developmental delay and history of focal epileptic seizures, presented to the MRI suite for a diagnostic scan. At about 1 month after normal birth, she was noted to have a short neck and restricted movements of shoulders and wrists. The mother gave a history of snoring during sleep with a protruded tongue. On examination, she had short stature with frontal bossing and short neck. She was uncooperative with regard to further airway manipulation, owing to the atlantoaxial instability.1,3 Unsuccessful intubation attempts have resulted in airway-related deaths of children having mucopolysaccharidoses.4 Dealing with such a challenging scenario at a remote location outside operating rooms is a trying ordeal for an anesthesiologist.

Discussion
Children with Hurler’s syndrome can present with abnormal upper airway anatomy, poor thoracic compliance, and atlantoaxial instability.1,3 Unsuccessful intubation attempts have also resulted in airway-related deaths of children having mucopolysaccharidoses.4 The difficult airway encountered in Hurler’s syndrome is attributed to the presence of macrocephaly, macroglossia, odontoid hypoplasia, dental distortions, gingival hyperplasia, swollen epiglottis, tonsillar enlargement, and anteriorization of the larynx.2,5 Face mask ventilation could be difficult and an adequate seal can be achieved with the help of an inverted mask technique.5 Narrowing of laryngeal and tracheobronchial

Abstract
Hurler’s syndrome is a rare genetic disease characterized by progressive multiorgan accumulation of glycosaminoglycans. It is associated with progressive craniofacial, skeletal, and cardiac involvement, which increases the risk of anesthesia. Patients with Hurler’s syndrome could present as the worst airway management problem an anesthesiologist could deal with due to abnormal upper airway anatomy and limited neck manipulation, owing to the atlantoaxial instability. We report a case of difficult airway scenario of a child with Hurler’s syndrome, leading to an apparent cannot ventilate cannot intubate scenario, which was managed successfully with the help of a C-MAC video laryngoscope with unique D-blade. In Hurler’s syndrome, C-MAC with D-blade is an excellent tool in establishing an airway in a pediatric difficult airway scenario. Moreover, D-blade C-MAC could be considered as the primary tool for establishing an airway in pediatric patients with Hurler’s syndrome.

Keywords
► Hurler’s syndrome
► pediatric difficult airway
► D-blade C-MAC
cartilage often necessitates the need for a smaller-sized ETT.\textsuperscript{1}
In our case, we secured the airway with a 3.5 size tube rather
than in accordance with the appropriate for age formula, due
to apparently smaller glottis which can be attributed to the
deposition of glycosaminoglycans in the airway. We were
able to ventilate without any leak. Due to the anticipated dif-
ficult airway in the majority of the patients, blind intubation
can be tried,\textsuperscript{1} but in our case, we were not able to secure the
airway with bougie-assisted blind oral intubation. Although
LMA is useful in an emergency scenario in establishing a suc-
cessful airway in some patients, it failed to guarantee ade-
quate ventilation in our patient. A case report by Busoni et al,
which described how LMA could lead to inspiratory stridor
and airway obstruction in mucopolysaccharidosis II (Hunter
syndrome), has been published, and these findings can be
extrapolated for our patient due to resemblance in airway
morphology.\textsuperscript{6} Hurler’s syndrome is known to cause thick
secretions in the airway and collapse of supraglottis after
anesthesia, which could have inhibited effective ventilation in
our patient at a later stage, even with an LMA.\textsuperscript{1}
In our scenario, we succeeded in securing the airway in
the first attempt with the C-MAC pediatric D-blade and the
head in the neutral position. Pediatric D-blade has been
specially designed for difficult airway with CL III or IV and
an acutely angulated design, giving it a half-moon shape.\textsuperscript{7}
The degree of angulation is 40° compared with 18° with a con-
ventional C-MAC Mackintosh blade. The neutral position of
the head is ideal due to the high-incidence of odontoid
dysplasia in this group of patients, which could be ensured
with the help of this unique video laryngoscope. Although
the DAS pediatric difficult airway guidelines advocate the
percutaneous cricothyroidotomy in cannot ventilate can-
not intubate scenario after repeated attempts with LMA,
C-MAC with a pediatric D-blade could be considered as a
noninvasive and appealing option for these patients.\textsuperscript{8}

\textbf{Conclusion}
In Hurler’s syndrome, C-MAC with D-blade is an excellent
tool in establishing an airway in a pediatric difficult airway
scenario. Moreover, D-blade C-MAC could be considered
as the primary tool for establishing an airway in pediatric
patients with Hurler’s syndrome.

\textbf{Conflict of Interest}
None declared.

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