

## Marshall-Smith Syndrome- Anaesthetic Considerations

Rakesh Garg, Gurpreet Singh, Maya Dehran

---

Marshall et al described a syndrome in 1971, characterized by accelerated osseous maturation, orofacial dysmorphism, mental retardation and failure to thrive.<sup>1,2</sup> Subsequently this syndrome was labeled as Marshall Smith syndrome. Significant respiratory problems are the main cause of death.<sup>3</sup> We are reporting a 3-year-old male child with this syndrome, who had suffered significant respiratory problems but was managed successfully to survive till this age as a result of timely airway intervention.

### CASE REPORT

3-year-old male child weighing 10 kg, with Marshall Smith syndrome was scheduled for tracheoscopy and assessment of upper airway.

He was born at term by caesarean section delivery and developed stridor on the first day of life which was managed conservatively. On day three, he developed cyanosis following a feed. Initially he was managed conservatively but subsequently he required tracheal intubation and mechanical ventilation. His old records revealed difficult intubation with anteriorly placed larynx. After 2 days, weaning was tried but was unsuccessful. Therefore, bronchoscopy was done which revealed features of moderate laryngomalacia and bilateral choanal atresia.

On further evaluation in the paediatric genetic unit, he was diagnosed to have Marshall Smith Syndrome. MRI scan of the cervical spine revealed a normal atlanto occipital joint, cervical vertebra, cervical spinal cord and cervical canal but the cerebellar tonsils were extending 6 mm beyond the lower border of occipital bone. Later, he was tracheostomised and was gradually weaned off the ventilator. He was discharged after 18 days. He continued to have recurrent chest infection and at the age of 7 months, another bronchoscopy was done which revealed laryngomalacia and subglottic stenosis.

On preanaesthetic check up, he had dysmorphic facial features with mid facial hypoplasia, depressed nasal bridge, bilateral proptosis, blue sclera, megalocornea, increased body hair, long toes and prominent heels. He had tracheostomy tube size 4.5 mm ID in situ. He was

premedicated with oral midazolam 5 mg, one hour prior in the preanaesthetic room. He was shifted to operation room and routine monitors including electrocardiogram (ECG), noninvasive blood pressure (NIBP), pulse oximeter were attached. Anaesthesia was induced with sevoflurane in oxygen and nitrous oxide via tracheostomy tube using Ayre's T piece circuit. A 22G intravenous access was secured, injection fentanyl 20 µg and succinylcholine 15 mg were administered. Flexible fiberoptic bronchoscopy revealed a membrane just below the glottis almost completely obscuring the lumen. Bronchoscopy was done via tracheostomy stoma to assess the airway below the membranous obstruction. Since bag and mask ventilation was not possible in view of the obstruction and dysmorphic features, 100% oxygen was administered before proceeding for tracheoscopy. Bronchoscopy confirmed the presence of membranous obstruction and tracheal stenosis. Tracheostomy tube was reinserted and ventilated with 100% oxygen till the child had adequate respiratory efforts. He had an uneventful recovery and was discharged the next day.

### DISCUSSION

The facial dysmorphism in children with Marshall Smith Syndrome includes prominent eyes and forehead, megalocornea, micrognathia and anteverted nostrils with small nose. The other anomalies occasionally reported are scoliosis, atlanto-occipital instability, choanal atresia, abnormal larynx or laryngomalacia, cardiovascular defects, hydronephrosis and immunological abnormalities.<sup>2,3,4</sup>

Children with Marshall Smith Syndrome are posted for repeated airway evaluation requiring anaesthesia which poses a challenge to anaesthesiologist in view of upper airway problems and possibility of atlanto-axial instability along with facial dysmorphism. They may cause difficulty in ventilation with face mask as well as in intubation. This respiratory problem can be functional obstruction of the upper airway but can also be due to structural tracheal problem and restrictive syndrome condition due to hypoplastic thorax. Ideally, spontaneous ventilation should be preserved during induction of anaesthesia until the airway

---

*Drs. Rakesh Garg, Gurpreet Singh, Senior Residents, Maya Dehran, Professor, Department of Anaesthesiology and Intensive Care, All India Institute of Medical Sciences, Ansari Nagar, New Delhi -110029, India*

*Correspondence : Dr Rakesh Garg, Email: drrgarg@hotmail.com*



**Figure 1**

*Child of Marshal Smith Syndrome*

is controlled. Functional problems at the level of the hypopharynx may be circumvented by the use of a nasopharyngeal airway.

Airway difficulty is described in the literature,<sup>5</sup> but this child had laryngeal web almost completely obscuring the glottic inlet. So the gold standard of fiberoptic intubation was not feasible. This child was earlier tracheostomized, and hence the difficulty in securing the airway was not of much concern. Tracheoscopy via stoma to evaluate was a concern as tracheostomy tube needed to be removed. We had kept randel backer saucek mask (for tracheostomy

stoma ventilation) connected with swivel connector ready to use while performing tracheoscopy, in case patient starts desaturating.

The tracheostomy helped in circumventing problems associated with difficult intubation due to an anteriorly placed larynx, functional problems of the upper airway and possible atlanto-occipital instability.

To conclude, children with Marshall Smith Syndrome requires great vigilance in the perioperative period in view of associated airway and respiratory problems.

## REFERENCES

1. Sharma AK, Haldar A, Phadke S, Agarwal SS. Marshall-Smith Syndrome: A distinct Entity. *Indian Pediatrics* 1994; 31: 1098-2001.
2. Charon A, Gillerot Y, Maldergem LV, schaftingen MHV, Bont BD, Koulischer L. The Marshall-Smith syndrome. *Eur J Pediatrics* 1990; 150: 54-55.
3. Antila H, Laitio T, Aantaa R, Silvonieminen P, Pakkanen A. Difficult airway in a patient with Marshall Smith Syndrome. *Pediatr Anaesth* 1998; 8: 429-432.
4. Eich GF, Silver MM, Weksberg R, Daneman A, Costa T. Marshall-Smith syndrome: new radiographic, clinical and pathologic observations. *Radiology* 1991; 181: 183-188.
5. Yoko W, Yoko T, Naoji U, Toshiya K. A case of marshall-smith syndrome. *Jpn J Anesthesiol* 2003; 52: 86-862.

## JOURNAL OF ANAESTHESIOLOGY CLINICAL PHARMACOLOGY

invites you to visit

[www.joacp.org](http://www.joacp.org)

*One site that gives to access to all you desire in Anaesthesiology  
& utilize the following features :*

- |                          |                    |                               |
|--------------------------|--------------------|-------------------------------|
| ◦ Guidelines for Authors | ◦ Subscribe        | ◦ Submit your Articles online |
| ◦ Current Issue          | ◦ Journal Archives | ◦ Conference Calendar         |
| ◦ Search                 | ◦ Downloads        | ◦ Be our Peer Reviewer        |
| ◦ Jobs/Vacancy           | ◦ News             | ◦ RSS Feeds                   |
| ◦ Forums                 | ◦ Societies        | ◦ Contact us                  |

### LINKS TO PROMINENT ANAESTHESIOLOGY JOURNALS

- |                                       |                                       |
|---------------------------------------|---------------------------------------|
| ❖ Indian Journal of Anaesthesia       | ❖ Indian Journal of Pharmacology      |
| ❖ Anaesthesia                         | ❖ Anesthesiology                      |
| ❖ Anesthesia & Analgesia              | ❖ British Journal of Anaesthesia      |
| ❖ Canadian Journal of Anaesthesia     | ❖ European Journal of Anaesthesiology |
| ❖ Pain                                | ❖ RAPM                                |
| ❖ Acta Anaesthesiologica Scandinavica | ❖ Resuscitation                       |
| ❖ Survey of Anesthesiology            | ❖ Pediatric Anesthesia                |
| ❖ Other Journals                      |                                       |

### GUIDELINES BY VARIOUS ANAESTHESIOLOGY SOCIETIES

- |                       |                         |                     |
|-----------------------|-------------------------|---------------------|
| ✘ ASA Guidelines      | ✘ CPR Guidelines        | ✘ AAGBI Guidelines  |
| ✘ PAIN Guidelines     | ✘ ASRA Guidelines       | ✘ TRAUMA Guidelines |
| ✘ CANADIAN Guidelines | ✘ COMPASS Guidelines    | ✘ ESRA Guidelines   |
| ✘ IDSA Guidelines     | ✘ OBS-ANESTH Guidelines | ✘ CCM Guidelines    |
| ✘ MSA Guidelines      | ✘ SCA Guidelines        |                     |