

# Successfully Anesthetic Management in a Rare Syndrome, Noonan Syndrome: Case Report



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**Submission:** January 29, 2017; **Published:** May 30, 2017

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## Abstract

Noonan's Syndrome (NS) is a multisystem congenital disorder and characterized by fascial and physical features along with congenital heart disease. In these patients, fascial features include short webbed neck, micrognathia, limited mouth opening and high arched palate can be a big problem for the tracheal intubation. Most commonly associated cardiac anomaly is pulmonary stenosis (>50% of cases). In these patients anesthetic management is important because of difficult airway and severe cardiac abnormalities. Here, we have reported the preoperative evaluation and anaesthetic management of a child with NS complicated by pulmonary stenosis and laringomalasia.

**Keywords:** Congenital heart surgery; Noonan's syndrome; Difficult intubation; Pulmonary stenosis

**Abbreviation:** NS: Noonan's Syndrome; PS: Pulmonary Stenosis; HOCM: Hypertrophic Obstructive Cardiomyopathy; ASD: Atrial Septal Defect; RVOTR: Right Ventricle Outflow Tract Reconstruction; BIS: Bispectral Index; FOB: Fiber-Optic Bronchoscopy; IOC: Index Of Consciousness

## Introduction

Noonan's Syndrome (NS) is a multisystem congenital disorder. Pulmonary stenosis (PS) and hypertrophic obstructive cardiomyopathy (HOCM) are the most common cardiac anomalies found in combination with other lesions like atrial septal defect (ASD), ventricular septal defect, tetralogy of Fallot, aortic stenosis, coarctation of aorta, Ebstein's malformation, total anomalous pulmonary venous return, ostium primum ASD and patent ductus arteriosus [1,2]. The potential anesthetic problems presented by a patient with Noonan's syndrome may be due to impairment of cardiopulmonary function, the possibility of a difficult airway [1,2].

In this case report we have described the preoperative evaluation and anaesthetic management of a child with NS complicated by pulmonary (PS) stenosis and laringomalasia. The anesthetic management in these patients should be carried out with careful preoperative evaluation of physical status, to maintain hemodynamic stability and especially the difficult endotracheal intubation should be kept in mind [3]. In this case, we have reported an anesthetic management of a 13 months old girl with Noonan Syndrome underwent congenital heart surgery under general anesthesia suggesting difficult tracheal intubation

on preoperative physical examination and evaluation of the anesthesiologic aspects of this syndrome.

## Case Report

A thirteen-months-old, 5kg weight girl with clinical diagnosis of NS was admitted to our hospital for the operation of PS. In echocardiography PS had detected at birth. In the third month balloon valvuloplasty was performed. Pulmonary gradient was 68mmHg. Because of growth deficiency RVOTR (Right Ventricle Outflow Tract Reconstruction) was planned for her.

In the preoperative evaluation she had pes plano valgus, wide hemangioma in face, short stature, micrognathia, macroglossia, short stature. She had gone to operation for PS under general anesthesia. Monitoring was done throughout the operation; vitals were recorded on monitors every 5 minutes. In addition to standard monitoring, sedation level monitored with Bispectral Index (BIS), regional oxygen saturation was monitored with INVOS Cerebral Oximeter, which afford dual-site monitoring with pediatric disposable sensor, invasive right arterial pressure also monitored. In case of intubation and airway management difficulties we prepared equipments, fiberoptic laryngoscopy for difficult intubation.

General anesthesia was initiated with 2mcg/kg/min remifentanyl infusion. After establishing successful bag mask ventilation Cormack-Lehane score 3 were assessed with direct laryngoscopy (macintosh blade, size 2). After two failed intubation attempts with direct laryngoscopy, fiber-optic bronchoscopy (FOB) was carried out and a successful intubation with a FOB was performed. In the meantime, no desaturation occurred in the patient who was ventilated with the mask. Cuffed endotracheal tube number 4 mm internal diameter was safely placed into the trachea without trouble.

Endotracheal cuff pressure was maintained among 10-15cm H<sub>2</sub>O which was continuously monitored till extubation. After confirming effective endotracheal intubation, sodium thiopental 3-4mg/kg and rocuronium 0.6mg/kg were given. Anesthesia was maintained with isoflurane 50% oxygen/air and continuous infusion of remifentanyl (0.01-1mcg/kg/min) until the end of surgical procedures with the aim of keeping index of consciousness (IOC) values within 40-60, heart rate and blood pressure within the 30% range. Depth of anesthesia was monitored with index of consciousness (IOC), Morpheus Medical, Barcelona, Spain [K]. Cerebral (rSO-C) and somatic (rSO-S) tissue oxygen saturation were monitored and were stable during the operation, did not change compared to the initial values.

Data were continuously updated at two readings per second and average recordings saved at 1 minute intervals (Pediatric SomaSensor, Model SPFB, for children 4-40kg by Somanetics Corporation, Troy, Michigan for the INVOS 5100 Cerebral oximeter). Remifentanyl, being an ultra short active opioid, was preferred for slow induction of anesthesia being advantageous for hemodynamic stability in this case. At the end of operation the patient was taken to the CICU; mechanical ventilation was done in SIMV mode. Patient was extubated when her spontaneous breathing was adequate 3 hours later after the operation without any problem.

## Discussion

Noonan syndrome (NS) with characteristic facial features is one of the most common non chromosomal syndromes presenting to the cardiac anesthesiologist for the management of various cardiac lesions, predominantly pulmonary stenosis (PS) (80%) and hypertrophic obstructive cardiomyopathy (HOCM) (30%) [4,5]. Anesthetic management poses a multitude of challenges, especially related to the airway management and maintenance of cardiovascular stability.

Successful management of the difficult airway implies recognition, adequate preparation, and, finally, familiarity with at least several of the special techniques which may be used for tracheal intubation of patients with a difficult airway [3]. Noonan syndrome is characterized by fascial and physical features, airway abnormalities along with congenital heart disease. In these patients, fascial features include short webbed neck, micrognathia, limited mouth opening and high arched palate. Pulmonary stenosis and hypertrophic

obstructive cardiomyopathy are highly prevalent. The anesthetic management is important because of difficult airway and severe cardiac abnormalities [3-6].

Here, we have reported a thirteen-months old patient with characteristic features of NS; micrognathia, limited mouth opening, high arched palate, short stature, macroglossia who underwent congenital heart surgery without any complication in airway management during the operation. Miscellaneous conditions such as macroglossia, micrognathia, short stature can congest small infant mediastinum compromising airway [3-6]. Since our patient had the characteristic features macroglossia, micrognathia, laryngomalacia; resuscitation and difficult airway management trolley, FOB was kept by the side. Equipment for tracheostomy was made available. We have used FOB for intubation after two inadequate attempts for intubation with direct laryngoscopy (macintosh blade, size 2).

The airway trolley was made ready with all the equipment necessary in case of difficult airway management. Arterial hypoxemia and desaturation can occur very rapidly in pediatric patients in view of the decreased functional residual capacity. Therefore, preoperative, intraoperative, and postoperative pharmacology has to be given an in-depth consideration to avoid any incidence of hypotension and hypoxemia [6-8].

After two failed intubation attempts with direct laryngoscopy, a successful intubation with a fiberoptic bronchoscope was performed. After operation the patient was transferred to CICU. We planned to apply fast track extubation that's why we used ultra-short acting opioid, remifentanyl during surgery and CICU.

When early extubation is planned, ensuring adequate postoperative pain management is essential. Endotracheal cuff (ETTc) pressure monitored till extubation between the ranges of 10-15cm H<sub>2</sub>O. The tracheal tube cuff should ideally seal the airway without compromising mucosal perfusion, cuff pressure should be maintained around 10-15cm H<sub>2</sub>O in critically ill intubated and mechanically ventilated patients. When ETTc pressure exceeds the capillary perfusion pressure of tracheal mucosa, mucosal blood flow is obstructed and may lead to severe even fatal injury including tracheal pain or stridor [9,10].

Early extubation after congenital heart surgery is becoming popular in selected patients. Fast-track cardiac anesthetic techniques lead to earlier tracheal extubation, shorter ICU stays and significant reductions in cost. Remifentanyl, ultra short acting opioid, has been used for infants and children to achieve easy, safe and early extubation after major surgeries [11-14]. Achieving adequate depth of anesthesia during surgical procedures is desirable. IOC can be useful in guiding anesthetic dose to avoid risks of intraoperative recall in surgical patients with high risk of awareness; can improve anesthetic delivery and recovery from anesthesia. We used IOC monitoring both perioperative and postoperative period to determine the depth of anesthesia and to maintain optimal conditions for extubation [15,16].

Successful management of the difficult pediatric airway can be challenging and stressful. Anticipation is often key to success, and it is preferable to err on the side of conservatism. In the unanticipated difficult airway, anesthesia personnel must utilize the conservative, «common sense» approach advocated in the ASA guidelines. Successful management of the difficult airway implies recognition, adequate preparation, and, finally, familiarity with at least several of the special techniques which may be used to intubate the trachea of patients with a difficult airway. For successful airway and anesthetic management in a case of NS, anesthesiologists should have thorough and deep knowledge about the various anatomic anomalies and pathophysiologic considerations to prevent any clinical disaster, especially for an elective surgery.

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DOI: [10.19080/JAICM.2017.02.555594](https://doi.org/10.19080/JAICM.2017.02.555594)

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