

## CASE REPORT

# Anesthetic management in a pediatric patient with Noonan syndrome and pulmonary stenosis: a case report

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### ABSTRACT

This case report describes anesthetic considerations in a 7 year old boy with Noonan syndrome posted for repair of unilateral undescended testis. The patient had pulmonary stenosis for which pulmonary balloon valvoplasty was done under general anesthesia at his 4 years of age. This affected anesthetic plan.

**Key words:** Anesthesia, General; Noonan syndrome; Pulmonary stenosis

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### INTRODUCTION

Noonan's syndrome was first described by Noonan and Ehmke in 1963.<sup>1,2</sup> It is a rare clinical entity and represents the phenotype of Turner's syndrome with normal chromosome studies.<sup>3</sup> These patients have facial, cardiovascular and skeletal abnormalities which may pose problems to the anesthetist during surgery.

Specific cause for Noonan syndrome is not known. It affects males and females equally. Inheritance is autosomal dominant but many cases seem to be sporadic.<sup>4</sup>

### CASE REPORT

A 7 year old boy, weighing 22 kg, was posted in surgery operation theatre for repair of left sided undescended testis. The diagnosis of Noonan syndrome was made when he was 4 years of age. Since early childhood he had frequent respiratory tract infection, cough and cold. The child had also history of delayed developmental milestones, palpitation and shortness of breath on exertion. He was undergone balloon pulmonary valvoplasty under general anesthesia at the age of 4 years for severe pulmonary stenosis. Thereafter, the child had normal growth with no significant medical

history. He was on oral tablet propranolol (10 mg/day). There was no significant family history for anesthetic complications or congenital abnormalities.

On examination, the child had hypertelorism, diversion of recti, pulse rate 92/min, blood pressure 110/64 mmHg, Mallampati score – grade 2, normal neck movement, adequate mouth opening. On auscultation, a systolic murmur was heard on left second intercostal space. The haemoglobin level was 12.8 gm/dl. Serum electrolytes, blood glucose, creatinine, rest of the investigations were normal. Hematological investigations showed no coagulations or platelet defects. Echocardiography showed mild pulmonary regurgitation.

On arrival in the operating room, pulse oximeter, electrocardiographic monitor, blood pressure cuff were attached. The child was cooperative. Peripheral venous access was secured using 22 G cannula. Antibiotic prophylaxis was given with IV 800 mg ampicillin and 24 mg gentamicin. Premedication was given with 2 mg midazolam, 0.1 mg glycopyrrolate, 50 µg fentanyl IV. Induction was done with 50 mg IV propofol. After establishing adequate bag mask ventilation, 2.2 mg IV vecuronium was given. After 3 min bag mask ventilation, intubation was done with 5.5 mm cuffed endotracheal tube.

Neuromuscular blockade was monitored using a peripheral nerve stimulator. Temperature was monitored using oesophageal probe. Maintenance of anesthesia was done with oxygen, nitrous oxide, sevoflurane, vecuronium bromide in the 1.5 hour procedure. 450 mg paracetamol infusion was given as analgesic. Ventilation was controlled to achieve an end-tidal CO<sub>2</sub> of 35-40 mmHg, and the SpO<sub>2</sub> was maintained between 95% and 100%. Total fluid deficit calculated was 434 ml, maintenance fluid 62 ml/hr. 550 ml lactated ringers solution was infused. At the end of the procedure, neuromuscular blockade was reversed with the intravenous administration of 0.2 mg of glycopyrrolate and 1.5 mg of neostigmine. Extubation was done after the return of rhythmic breathing and return of protective airway reflexes. The child was given postoperative oxygenation for 5 min and kept in postanesthesia care unit and careful monitoring was done for 24 hours. Diclofenac sodium was given as postoperative analgesic in infusion. Postoperative period was uneventful. The child was discharged.

## DISCUSSION

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 and the problem of technical difficulty with regional anesthesia. Noonan's syndrome is the XX or XY phenotype of Turner's syndrome, and is a distinct clinical entity.<sup>5,6</sup> In Noonan's syndrome 30-50% of the patients have pulmonary stenosis.<sup>7</sup> Hypertrophic obstructive cardiomyopathy, atrial septal defects, patent ductus arteriosus may also be associated with

Noonan syndrome.<sup>8,9</sup> Other various associations of this syndrome are hypertelorism, downward eye slant, epicanthal fold, ptosis, flat nasal bridge, high palatal arch, dental malocclusion, micrognathia, short webbed neck, short stature, bleeding diathesis, deafness, developmental delay, mild mental retardation, hepatosplenomegaly, scoliosis. Patients may exhibit cubitum valgum, clinodactyly, and vertebral anomalies that may affect positioning. Protection of pressure points with padding during intraoperative period is important.

M. Lynn et al, in a study in 2007, described anesthetic management of a 6 year old boy with Noonan syndrome having mastocytosis, von Willebrand disease, patent foramen ovale, pulmonary stenosis posted for adenoidectomy. The patient's induction, maintenance, emergence were uneventful.

Our patient was postoperative patient for pulmonary stenosis. He had hypertelorism, diversion of recti but not associated with difficult airway, coagulopathy. Antibiotic prophylaxis with IV injection ampicillin and gentamicin was given. Avoidance of tachycardia due to sympathetic stimulation and stress response, hypotension, maintenance of preload and afterload were important considerations. Beta-adrenergic blockade with propranolol was continued. Intraoperative and postoperative periods were uneventful. In postoperative period, factors precipitating pain, hypotension, hypovolemia, hypothermia and sympathetic stimulation were avoided as these were detrimental to cardio-pulmonary system.

**Conflict of interest:** None declared by the authors.

**Author contribution:** All authors took part in the management of the patient and preparation of the manuscript.

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