Anaesthesia for emergency ventriculo-peritoneal shunt in an adolescent with Noonan's syndrome

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See commentary "Treatment of hydrocephalus: Challenges and the way ahead" on page 456.

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Abstract

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A 15-year-old boy with Noonan's syndrome was admitted for emergency ventriculo-peritoneal shunt. Intraoperative course was complicated by hypertensive urgency, which was effectively managed with high doses of esmolol (500 μg/kg/min). Difficult airway was anticipated due to presence of webbed neck and facial dysmorphism. Tracheal intubation was however successfully accomplished with the aid of a bougie. This report thus highlights the unique anaesthetic problems encountered during anaesthetic management of such a case, which is worth sharing.

Keywords: Hypertension, hypertrophic cardiomyopathy, Noonan syndrome, paediatric anaesthesia, scoliosis

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INTRODUCTION

Noonan's syndrome (NS) is an autosomal dominant congenital disorder that affects both males and females equally.[1] It is characterised by short stature, facial dysmorphology and congenital heart defects. webbed neck, chest deformity, mild intellectual deficit, cryptorchidism, bleeding tendency and lymphatic dysplasias are some of the other associated deformities. The severity of clinical features can vary from mild to severe and thus the syndrome may be missed at an early age.

We present the anaesthetic management of a 15-year-old short statured boy with hydrocephalus scheduled for an emergency ventriculoperitoneal (VP) shunt.

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CASE REPORT

A 15-year-old adolescent presented to the neurosurgical emergency department with history of blurring of vision for past 15 days. He was drowsy and his Glasgow coma scale (GCS) was 12/15. He was referred from a private health care centre where he was being managed for

tubercular meningitis for past 1-month.

The patient was short statured (140 cm) and weighed 25 kg. His facial abnormalities included hypertelorism, downslanting prominent eyes, thick lips and high palatal arch. His Mallampati score was 3, neck extension was limited but mouth opening was 4 cm. Scoliosis of dorsal spine, reduced volume of thorax, systolic murmur at left sternal edge, sinus tachycardia (heart rate [HR] =140 beats/min) and systemic hypertension (blood pressure [BP] =180/100 mm of Hg) were the positive findings on clinical examination. His high BP had been detected during his previous hospitalization but was not treated with any antihypertensives as it was considered to be secondary to intracranial hypertension (Cushings reflex).

None of his family members had features suggestive of NS. The boy had no past history of any medical or surgical intervention. The father however, observed mild motor delay, growth retardation and physical deformities in the boy since birth. Presently, the patient had no symptoms pointing toward any cardiac or respiratory decompensation.

The haemoglobin level was 12.8 g/dl, and the serum electrolytes, glucose, liver and renal function tests, coagulation profile (prothrombin time and partial thromboplastin time) and platelet counts were within the normal limits. Chest X-ray [Figure 1a], X-ray neck anterior-posterior and lateral view [Figure 1b], non-contrast computed tomography head and magnetic resonance imaging of the brain were performed. Basilar invagination and atlantoaxial dislocation could not be ruled out in X-ray neck due to poor visualisation of atlas and axis and altered alignment of the cervical spine. Sinus tachycardia, left ventricular hypertrophy and Q-waves in lead II, III, aVF, V5 and V6 were seen on electrocardiogram. Transthoracic echocardiography was suggestive of hypertrophic cardiomyopathy (HCM) with no left ventricular outflow tract (LVOT) obstruction and no evidence of systolic anterior motion of the mitral valve. Details could not be assessed in view of poor acoustic window due to marked dorsal scoliosis and mal alignment of the ribs.

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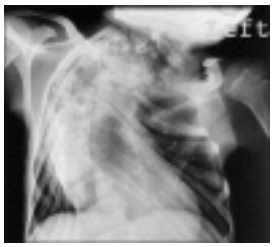

Figure 1a

caption a4

Chest X-ray. (i) Scoliotic deformity of dorsal spine (Cobbs angle 50), (ii) vertebral bodies rotated, malalignment of ribs, (iii) reduced volume of thorax, (iv) abnormal curvature of tracheomediastinum, (v) normal calibre of trachea, (vi) lung fields **...**

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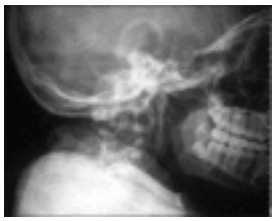

Figure 1b

caption a4

X-ray neck (lateral view)

Standard monitors were attached in the operation theatre and HR of 130 beats per min, BP of 180/100 mm Hg and SPO2 of 100% was recorded. Antibiotic prophylaxis was administered (cefuroxime 50 mg/kg initial dose followed by twice daily dose). Resuscitation and difficult airway cart (e.g. supraglottic airway devices and fibre optic bronchoscope) was kept ready. The child was premedicated with intravenous (IV) midazolam (1.5 mg) and right radial artery was cannulated for continuous monitoring of BP prior to induction. The child had high BP; systolic and diastolic pressures were in the range of 180-220 and 100-120 mm Hg respectively. Esmolol was administered twice in a bolus dose of 10 mg to control the high BP. A decrease of 25% from the baseline recordings was targeted and esmolol infusion at the rate of 500 μg/kg/min was administered to maintain the same. Induction of anaesthesia was carried out after stabilisation of systolic pressures at 140-160 mm Hg.

After preoxygenation for 5 min, patient was induced with 3 mg of morphine and thiopentone (100 mg). After ensuring successful bag mask ventilation, check laryngoscopy was performed and Cormack Lehane Grade 2b of glottic view was obtained with use of backward, upward, and rightward pressure manoeuvre. Vecuronium bromide (2.5 mg) was administered and fentanyl (40 μg) and lignocaine (30 mg) were administered to blunt pressor response to laryngoscopy and intubation. Trachea was intubated by rail roading a 6.5 mm cuffed endotracheal tube over a bougie. Left internal jugular vein was cannulated for central venous monitoring; surgeons had planned a right VP shunt and thus this side had to be avoided.

Anaesthesia was maintained with O2/N2O and sevoflurane (1.5 minimum alveolar concentration) and small tidal volumes (4 ml/kg) and high respiratory rates were used to maintain sufficient minute ventilation. End tidal CO2 concentration was being continuously monitored and maintained at 30 mm of Hg. The estimated blood loss was 50 ml, duration of surgery was 120 min and results of intraoperative blood gas analysis were normal; pH 7.5, PaO2 97 mm Hg, PaCO2 29 mm Hg, and HCO3 of 22 mmol/L. Urine output and temperature were also monitored. Reversal of residual paralysis was accomplished with IV administration of 0.2 mg of glycopyrrolate and 1.5 mg of neostigmine and trachea was successfully extubated. His GCS was 12 and he was transferred to Intensive Care Unit (ICU) for haemodynamic and neurological monitoring. Unfortunately, he developed shunt obstruction, his GCS deteriorated

(GCS = 7) and he was re-intubated the next day. He subsequently died of septic shock and multi organ dysfunction syndrome after 15 days of stay in the ICU.

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DISCUSSION

Exact cause of hypertension in our patient could not be elucidated; whether neurogenic, co-existing essential hypertension or secondary to renal abnormalities associated with NS. There is only one case report of a child with NS and hypertension.[2] Vasodilators like nitroglycerin or nitroprusside are contraindicated in HCM; they decrease preload and systemic vascular resistance (SVR) and increase LVOT obstruction. They are also contraindicated in patients with elevated intracranial pressures.

Cardiovascular decompensation secondary to HCM was prevented by maintaining preload and afterload and avoiding any direct or reflex increase in contractility or HR.[3,4] Propofol decreases SVR and venous return whereas ketamine causes hypertension and tachycardia and both the agents are avoided for induction of anaesthesia. Opioids decrease the sympathetic and somatic responses to noxious stimulation and are advocated in patients with impaired cardiac function. Isoflurane and desflurane when administered in high concentrations increase HR and BP and are thus avoided. Halothane is advantageous in HCM in view of its negative inotropic action and maintenance of SVR. But we avoided its use in our case because it is also a cerebral vasodilator (halothane >>enflurane > desflurane > isoflurane > sevoflurane). Sevoflurane was preferred in our patient; it causes a modest decrease in SVR and BP with minimal action on cerebral blood flow. Complications such as congestive heart failure, myocardial ischaemia, systemic hypotension, and supraventricular or ventricular arrhythmias can be precipitated in patients with HCM secondary to dynamic LVOT obstruction and diastolic dysfunction.

High palatal arch, dental malocclusion, webbed neck and atlantoaxial instability complicate the airway management in patients with NS.[5] Though difficult airway was anticipated in our patient, intubation could be successfully accomplished with a bougie.

Associated anomalies such as pectus excavatum, kyphoscoliosis and short stature can cause respiratory compromise and need for post-operative mechanical ventilation. Vertebral anomalies can pose technical difficulties with regional anaesthesia.[5]

Factor VIII, XI, XII deficiencies, thrombocytopenia and platelet function defects can cause increased bleeding.[6] Suitable blood products should be kept ready so that intraoperative and post-operative haemorrhage can be managed.

Williams, Aarskog, Cardio-facio-cutaneous syndrome, Costello, LEOPARD and King-Denborough syndrome are some of the differential diagnoses.[7] 7Diagnosis of Noonan's syndrome is based on Van der Burgt criteria [Table 1].[8] Phenotypic variability in Noonan's syndrome leads to its underdiagnosis.

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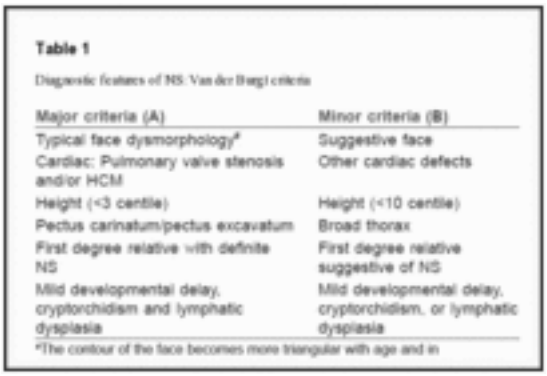

Table 1

Diagnostic features of NS: Van der Burgt criteria

Major criteria (A)	Minor criteria (B)
Typical face dysmorphology[a]	Suggestive face
Cardiac: Pulmonary valve stenosis and/or HCM	Other cardiac defects
Height (<3 centile)	Height (<10 centile)
Pectus carinatum/pectus excavatum	Broad thorax
First degree relative with definite NS	First degree relative suggestive of NS
Mild developmental delay, cryptorchidism and lymphatic dysplasia	Mild developmental delay, cryptorchidism, or lymphatic dysplasia

[a]The contour of the face becomes more triangular with age and in

Diagnostic features of NS: Van der Burgt criteriaGo to:

CONCLUSION

Children or adolescents with facial dysmorphology, short stature and cardiac defects should be evaluated for this syndrome. Anaesthetic management is planned on the basis of associated cardiac anomalies. Intense monitoring should continue in post operative period and surgical and anaesthetic adverse effects anticipated for a successful outcome.

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Footnotes

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Source of Support: Nil

Conflict of Interest: None declared

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