Anesthetic management of a patient with Meckle-Gruber Syndrome with complex cardiac anomalies for non-cardiac surgery

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Citation

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Abstract

An 8-month old male patient of Meckle-Gruber Syndrome with a single ventricle with unrestricted pulmonary blood flow leading, moderate ventriculoseptal defect, rudimentary right ventricle and a single atrioventricular valve presented for excision of occipital meningo-encephalocele. Previous banding of pulmonary artery trunk to reduce pulmonary blood flow had failed. Pre-anesthetic examination showed the patient to have pulmonary congestion secondary to excessive blood flow. General anesthesia with judicious use of hyperventilation, nitrous oxide and fluid restriction helped in diverting excess blood flow away from the pulmonary bed and prevented any further deterioration in pulmonary congestion. The strategy helped in a successful outcome from anesthesia in this patient having complex cardiac anomaly coming for non-cardiac surgery.

INTRODUCTION

Meckle-Gruber Syndrome [MKS] is a rare autosomal recessive disorder with an estimated incidence range of 1: 9000 [1] to 1:135, 0002. It is associated with occipital encephalocele, hypoplastic kidneys, polydactyly, cleft lip or palate, mandibular micrognathism, anatomical abnormality of the larynx and the tongue, and several other associated malformations including cardiac defects [3] . These patients usually die shortly after birth; those who survive have less severe deformities [4] . Recently we had an 8-month old patient of MKS with multisystemic disorder including a single cardiac ventricle. There is no literature describing the anesthetic management of patients with this rare condition of a single ventricle coming for non-cardiac surgery.

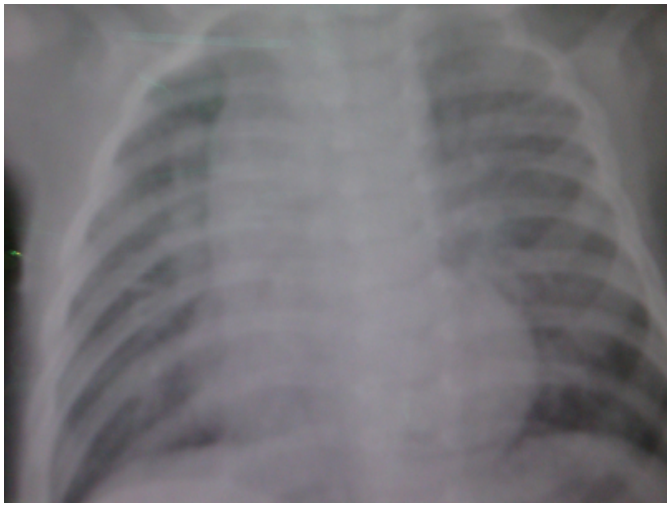
CASE REPORT

A 8-month old male patient weighing 5.5 kg with MKS was admitted for excision of occipital meningo-encephalocele. The patient had a complex congenital heart disease consisting of a single ventricle with unrestricted pulmonary blood flow, moderate ventriculoseptal defect, rudimentary right ventricle and a single atrioventricular valve. In addition, the patient had meningo-encephalocele, left cerebral leukomalacia and polycystic kidney. The patient had undergone pulmonary artery banding and balloon septostomy between 2 and 4 months after birth to reduce pulmonary blood flow. Post-banding systemic arterial saturation [SaO_2] was an acceptable 85-86% [5] .

Current pre-anesthetic examination showed the patient to have a SaO_2 of 95-96%. This was suggestive of pulmonary artery banding failure and a need for re-operation to tighten the pulmonary trunk band further. However the patient's parents insisted for excision of occipital meningo-encephalocele without any more cardiac surgery. Chest X-ray showed enlarged cardiac shadow with some lung congestion but no evidence of pleural effusion or pulmonary edema [Figure 1]. Vitals were stable. No airway problems were noted.

Figure 1

Figure 1: showing enlarged cardiac shadow with some lung congestion.



The patient was administered chemoprophylaxis for endocarditis and received syrup midazolam per orally as premedication. Anesthesia was induced with sevoflurane in oxygen. After IV cannulation, patient received 20 µg fentanyl and 5 mg atracurium. Following uneventful tracheal intubation, anesthesia was maintained on 60% nitrous oxide in oxygen with sevoflurane adjusted to maintain a blood pressure of ±20% pre-induction value. Tidal volume was gradually increased to 12 ml. kg $_{1}$ so as to induce stretching of pulmonary vasculature to increase pulmonary vascular resistance. Respiratory rate was adjusted to maintain end tidal carbon dioxide [$EtCO_2$] around 33-34 mmHg. With these ventilatory adjustments and a FiO2 of 0.4, oxygen saturation ranged between 91-94%. In addition to routine monitoring, invasive arterial and central venous pressures [CVP] were monitored. Intraoperatively, care was taken to maintain CVP between 4-6 cmH_2O in an attempt to reduce the chances of increasing pulmonary congestion.

During the entire duration of surgery [nearly 2 hours] patient maintained stable heart rate and blood pressure. No further deterioration was noted in lung congestion. He made an uneventful recovery in the postoperative period and was discharged 7 days after the surgery.

DISCUSSION

Cardiac repair and neurosurgical intervention is usually warranted in patients of MKS. Miyazu et al in 2005 $_{4}$ described a case of 2-year old patient with MKS necessitating general anesthesia for cardioplasty and gastrostomy. However their patient had no cardiac or nervous system lesions. The present patient posed anesthetic challenges due to his complex cardiac lesion and failure of pulmonary trunk banding.

The present patient underwent pulmonary artery banding at the age of 2 months as a palliative procedure to reduce excessive pulmonary blood flow. When correctly performed, this procedure produces fall in SaO_2. In general, SaO_2 between 75-85% is considered acceptable end point of successful banding $_{5}$. However, in our patient, somewhere over the next 6 months after initial banding, band failure occurred as indicated by the increased SaO_2 of 95-96%. Keeping this in mind and the complex cardiac pathology, we were forced to adopt a series of steps for successful outcome of anesthetic management.

First, we restricted intraoperative fluid administration to prevent any further increase in pulmonary congestion. This was done by intraoperative fluid restriction and keeping the central venous pressure between 4-6 cm H_2O. However the exact role of CVP monitoring in critically ill patient is unreliable $_{6}$. Pulmonary artery wedge pressures were not recorded as it was considered hazardous due to previous pulmonary trunk banding.

Second, hyperventilation produces vascular stretching and is known to reduce the pulmonary blood flow by increasing pulmonary vascular resistance $_{7}$. Our goal was to increase the pulmonary vascular resistance by hyperventilation. This promoted right to left shunt as evident by fall of SaO_2 between 91-94%. With this degree of hyperventilation $EtCO_2$ remained around 33-34 mmHg. We did not employ the technique of permissive hypercapnia to induce pulmonary vasoconstriction keeping in mind the possibility of inducing arrhythmias in the setting of low SaO_2.

Third, nitrous oxide was included in the anesthetic regime for its pulmonary vasoconstrictor effect $_{7}$ as this would increase pulmonary vascular resistance which was considered beneficial in this patient.

Lastly, advanced airway equipments for managing unanticipated difficult tracheal intubation were kept ready as these cases are known to pose difficulty in tracheal intubation $_{4}$.

In conclusion, this case report highlights several challenging handicaps in a patient of MKS with complex cardiac anomalies presenting for non-cardiac surgery which necessitated several corrective anesthetic strategies for a

successful outcome.

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References

1. Ahdab-Barmada M, Claassen D. A distinctive triad of malformations of the central nervoue system in the Meckle-Gruber syndrome. J Neuropathol Exp Neurol 1990; 49: 610-620.
2. Auber B, Burfeind P, Herold S, Schoner K, Simson G, Rauskolb R, Rehder H. A disease causing deletion of 29 base pairs in intron 15 in the MKS1 gene is highly associated with the campomelic variant of the Meckle-Gruber syndrome. Clin Genet 2007; 72: 454-459.
3. Frank V, den Hoallander AI, Bruchle NO, Zonneveld MN, Nurnberg G, Becker C, Du Bois G, Kendziorra H, Roosing S, Senderek J, Nurnberg P, Cremers FP, Zerres K, Bergmann C. Mutations of the CEP290 gene encoding a centrosomal protein cause Meckel-Gruber syndrome. Hum Mutat 2008; 29: 45-52.
4. Miyazu M, Sobue K, Ito H, Azami T, Ito S, Takeuchi A, Sasano H, Tsuda T, Katsuya H. Anesthetic and airway management of general anesthesia in a patient with Meckle-Gruber Syndrome. J Anesth 2005; 19: 309-310.
5. Kouchoukos NT, Blackstone EH, Doty DB, Hanley FL, Karp RB. Cardiac Surgery, 3rd edn, Vol 2. Philadelphia: Churchill Livingstone; 2003. p.1114-1130.
6. Vincent JL, Weil MH. Fluid challenge revisited. Crit Care Med 2006; 34: 1333-1337.
7. Stoelting RK, Dierdorf SF. Anesthesia and co-existing diseases, 4th edn. Philadelphia: Churchill Livingstone; 2002. p.127-133.

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