

Anaesthesia Management in a Case of Walker Warburg Syndrome-A Case Report

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Abstract

Walker Warburg Syndrome is extremely rare autosomal recessive disorder with a reported incidence of 1.2 per 100,000 live births. Typical findings include hydrocephalus, type II cobblestone lissencephaly, cerebellar malformation, retinal dysplasia and muscular dystrophy. We report the anaesthetic management of a 21 days old child with Walker Warburg Syndrome for ventriculo-peritoneal shunt insertion under general anaesthesia.

Keywords: Walker Warburg Syndrome; Anaesthesia management

Introduction

Walker Warburg Syndrome (WWS), also called cerebro-ocular dysplasia-muscular dystrophy syndrome, is a rare form of autosomal recessive congenital muscle dystrophy [1]. Its incidence is 1.2 per 100,000 live births. It is the most serious form of congenital muscular atrophy with most patients succumbing before three years of age [2]. Many genes have been implicated in its etiology. Laboratory investigations usually show elevated creatine kinase, dysmorphic muscle pathology and altered alpha dystroglycan. The diagnostic criteria for WWS have been established by Dobyns et al. [3] and include type II lissencephaly, cerebellar malformation and ocular anomalies. Other brain anomalies such as, occipital encephalocele, absent corpus callosum, and fusion of the hemispheres are also reported in some cases. Besides these anomalies, cleft lip, cleft palate may be present in these children [4]. Delayed milestones, mental retardation and occasional seizures are other features of this syndrome. Cause of death in them is usually due to respiratory, failure, pneumonia, seizures, hyperthermia or ventricular fibrillation.

Owing to multisystem involvement, anaesthetic management of WWS present many challenges to anaesthesiologist. English literature search revealed only one case report of anaesthesia in WWS [5]. Here we present a case report 20 days old child with WWS who was brought to the neurocenter of our hospital and was diagnosed to have hydrocephalus which needed ventriculo-peritoneal shunt insertion.

Case Report

A twenty one days old full term vaginally delivered new born male weighing 3.5 kg, was referred to the neurocenter of our hospital from a peripheral hospital because of dysmorphic features. On examination the patient had receding mandible, brachycephally, short neck, widely spaced eye balls, bilateral exophthalmos with corneal opacities. On examination he was lethargic, hypotonic, with diminished reflexes and decreased spontaneous limb movements. He had weak sucking reflex. His cardiac and pulmonary systems were normal on auscultation.

His routine biochemical investigations including, blood glucose, serum electrolytes, urea and creatinine, were within normal range. His haemoglobin was 10.8 gm/dL. Investigations for inborn error of metabolism, congenital infections and chromosomal anomalies did not reveal anything significant. Brain MRI showed ventriculomegaly, thin corpus callosum and cerebellar hypoplasia, nearly complete agyria and irregular grey white matter junction, a pattern characteristic of cobblestone lissencephaly (Figure 1) Computerized tomogram of abdomen showed large cyst in right kidney. X-ray chest and

echocardiogram were normal.

He was admitted to neonatal intensive care unit (NICU) and in view of weak sucking reflex a nasogastric tube was inserted for feeding, and a 24 G cannula inserted into a foot vein. The child was kept fasting for 4 hours and infusion of 5% dextrose and half strength normal saline was started in the NICU. No premedication was given.

On arrival in the operation room, routine monitors (ECG, pulse oximeter and non invasive blood pressure) were attached. Difficult airway as well as malignant hyperthermia carts was kept in readiness. After doing gastric suction, nasogastric tube was removed. Anaesthesia was induced with fentanyl 6 mcg, propofol 15 mg, and tracheal intubation attempted without any muscle relaxant. Laryngoscopy (with number 1 Miller straight blade) revealed a view of Cormack Lehane Grade 2B (only the arytenoids and epiglottis seen). A size 3.5 endotracheal could be inserted on second attempt and confirmed by end tidal carbon dioxide and bilateral equal air entry on auscultation of lungs. Additional monitoring after intubation consisted of end

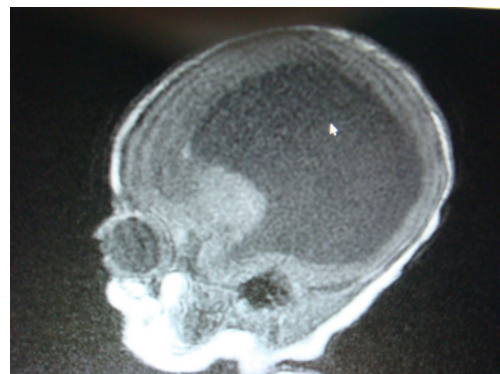


Figure 1: Brain MRI showed ventriculomegaly, thin corpus callosum and cerebellar hypoplasia, nearly complete agyria and irregular grey white matter junction, a pattern characteristic of cobblestone lissencephaly

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Received February 28, 2013; Accepted April 26, 2013; Published April 30, 2013

Citation: Mir GM (2013) Anaesthesia Management in a Case of Walker Warburg Syndrome-A Case Report. J Anesth Clin Res 4: 305. doi:10.4172/2155-6148.1000305

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tidal carbon dioxide, airway pressure, nasopharyngeal temperature and urine output. Baer Hugger warming system with an under body warming blanket was used to maintain normothermia. Anaesthesia was maintained with 50% oxygen in air, sevoflurane (inspired concentration 2-2.5%) and rocuronium one mg. Lungs were ventilated mechanically (pressure mode) to maintain end tidal carbon dioxide between 30 and 35 mmHg. The uneventful surgical procedure lasted 2 hours and blood loss was approximately 10 ml. At the end of surgery residual effect of muscle relaxant was not reversed and the patient was electively ventilated in the NICU where nasogastric tube was reinserted and trachea extubated after five hours. The child had smooth recovery and was transferred to ward next day.

Discussion

Children of WWS present many potential challenges to an anaesthesiologist, the most serious ones being difficult airway because of receding mandible, risk of aspiration as a consequence of pathology of gastrointestinal smooth muscles resulting in hypomotility of the intestinal tract. The problem is compounded by delayed gastric emptying which in conjunction with impaired swallowing and laryngeal reflexes increases the risk of postoperative pulmonary aspiration. They are also prone to development of malignant hyperthermia. Moreover, impaired renal functions, which may also accompany this syndrome, further complicate the anaesthesia management.

In anticipation of difficult intubation we kept ready difficult airway cart and did not administer any muscle relaxant to facilitate tracheal intubation, despite ease of mask ventilation. There is every likelihood of difficult laryngoscopy and intubation due to receding mandible. Repeated laryngoscopy and attempts at intubation may result in airway trauma and edema culminating in dangerous situation of cannot ventilate and cannot intubate [6]. Furthermore, suxamethonium administration in patients of muscular dystrophy has been known to result in dangerous hyperkalemia and rhabdomyolysis. Therefore, we avoided suxamethonium and administered rocuronium, that too, only after intubation.

Muscular dystrophy patients are prone to malignant hyperpyrexia which can be triggered by all the available volatile anaesthetic agents though; it has been reported infrequently with desflurane [7]. Therefore, one should anticipate and be prepared to manage this complication. Total intravenous anaesthesia with propofol has been found to be feasible and safe in children even less than 3 months of age [8], and has been recommended as anaesthesia technique of choice in patients predisposed to developing malignant hyperpyrexia. However,

we were handicapped by non availability of paedofusor TCI system for propofol infusion which is Target-Controlled Infusion (TCI) device that offers a means for producing relatively stable, controllable plasma concentrations of drugs administered intravenously. Due to associated genitourinary anomalies, these patients may have pre-existing renal dysfunctions with their own anaesthetic implications. Our patient was detected to have renal cyst but his renal functions were normal. In view of renal anomalies intraoperative urine output monitoring is highly imperative. Hence, measures must be taken to maintain normovolemia by infusing appropriate fluids and transfusing blood, if necessary. The fluid of choice, in a newborn undergoing neurosurgical procedure, should be dextrose in combination with half saline, because this combination, in addition to preventing hypoglycaemia will prevent brain edema because of its higher osmolarity compared to plasma.

Presence of muscle weakness may result in postoperative respiratory failure. These patients are at greater risk of postoperative aspiration of gastric contents due to impaired swallowing and delayed gastric emptying. Therefore, it is prudent to ventilate them during the early postoperative period [9].

Conclusion

We describe successful anaesthetic management of WWS with many associated anomalies which pose many challenges to anaesthesiologist.

References

1. Vajsar J, Schachter H (2006) Walker-Warburg syndrome. *Orphanet J Rare Dis* 1: 29.
2. Jones KL (1997) Walker Warburg Syndrome. *Fletcher J Smith's Patterns of Human Malformation*. WB Saunders Company, Philadelphia, USA 192-193.
3. Dobyns WB, Pagon RA, Armstrong D, Curry CJ, Greenberg F, et al. (1989) Diagnostic criteria for Walker-Warburg syndrome. *Am J Med Genet* 32: 195-210.
4. Burton BK, Dillard RG, Weaver RG (1987) Walker-Warburg syndrome with cleft lip and cleft palate in two sibs. *Am J Med Genet* 27: 537-541.
5. Sahajananda H, Meneges J (2003) Anaesthesia for a child with Walker-Warburg syndrome. *Paediatr Anaesth* 13: 624-628.
6. Curtis R, Lomax S, Patel B (2012) Use of sugammadex in a 'can't intubate, can't ventilate' situation. *Br J Anaesth* 108: 612-614.
7. Allen GC, Brubaker CL (1998) Human malignant hyperthermia associated with desflurane anesthesia. *Anesth Analg* 86: 1328-1331.
8. Steur RJ, Perez RS, De Lange JJ (2004) Dosage scheme for propofol in children under 3 years of age. *Paediatr Anaesth* 14: 462-467.
9. Barash PG, Cullen BF, Stoelting RK *Anesthesia for patients with rare and co-existing Diseases*. *Clinical Anesthesia* 505.