Anaesthesia Management of a New-born Baby with Walker-Warburg Syndrome undergoing Ventriculoperitoneal Shunt Insertion

Sir,

Walker-Warburg syndrome (WWS) is the most severe type of congenital muscular dystrophy (CMD) with very high mortality in early years of life.[1] Signs and symptoms include generalised decrease in muscle tone, muscle weakness, mental retardation, seizures and eye anomalies. Additionally, patients also have abnormalities in brain, like migrational defects with type II lissencephaly, hydrocephalus, cerebellar malformations and flat brainstem.[1] Laboratory findings include raised serum creatinine kinase levels, myopathic or dystrophic muscle pathology and abnormal alpha-dystroglycan. Management of these patients is only supportive with prevention of complications.[1] These patients are usually started on anticonvulsants if they develop seizures. Some children need neurosurgical interventions like shunting of hydrocephalus or encephalocele. Physiotherapy is needed to avoid development or worsening of muscle contractures. Some of these patients also need supplemental nasogastric or gastric tube feeding.[1]

We present a successful anaesthesia management to a six-day baby with WWS for the insertion of ventriculoperitoneal shunt to relieve hydrocephalus. The baby was delivered at full term by caesarean section and was transferred to neonatal intensive care unit (NICU) for trial of non-invasive ventilation due to poor respiratory efforts. On examination, he had generalised diminished reflexes. Chest auscultation revealed pansystolic murmur. Echocardiography showed mild to moderate patent ductus arteriosus (PDA). Laboratory investigations including full blood count, electrolytes and urea and creatinine were normal except high creatinine kinase (CK) and low magnesium levels. MRI brain showed large posterior fossa cyst communicating with fourth ventricle, cerebellar atrophy, dilated third and lateral ventricles, absence of septum pellucidum, absence of cerebral sulcation and microphthalamia of left eye. Kidney ultrasound revealed bilateral renal pelvic dilatation. On the basis of clinical and MRI findings with high CK levels, the diagnosis of WWS was made, which was confirmed with chromosomal analysis and muscle biopsy. His respiratory functions improved and magnesium levels became normal during his stay in NICU.

Perioperative management of patients with WWS is challenging, but has not been reported much in the anaesthesia literature. A detailed knowledge of the patients's medical condition is essential for adequate planning of these patients. WWS patients usually present with severe neurological impairment and muscle hypotonia making them prone to central and obstructive apnea. Preoperative sedation is, therefore, usually avoided. These patients might have reduced gastrointestinal motility and hence are more at risk for pulmonary aspiration.[2,3]

On arrival at operation room, routine monitors (ECG, pulse oximeter, and non-invasive blood pressure monitoring) were attached to the patient. Anaesthesia was induced with remifentanyl and propofol target control infusion (TCI) using Syramed^{Tm}µSP6000 at a blood concentration 4 µg/ml and 3-4 ng/ml, respectively. There is an association with potentially difficult airway[3] in these patients and that is why, we used glidoscope in our case. Considering the risk of rhabdomyolysis or non-specific hypermetabolic responses in patients with myopathies,[4] it is safe to avoid volatiles and succinylcholine in these patients and, hence, we used TCI propofol and remifentanyl at blood concentrations of 4 µg/ml and 3-4 ng/ml, respectively, for maintenance as well. Patient was meticulously monitored for any signs of malignant hyperthermia or rhabdomyolysis such as tachycardia, hyperthermia, muscle rigidity or marked increase in end tidal carbon dioxide. Intravenous (IV) dexamethasone, 0.3 mg, and paracetamol, 40 mg suppositories, were given. No muscle relaxant was used during the operation. Patient received a total 10 ml of 10% dextrose half saline as maintenance fluid intraoperatively. The operation lasted approximately two hours and was uneventful.

These patients are prone to develop respiratory complications in postoperative period because of poor respiratory efforts.[1] They should be transferred to intensive care unit postoperatively and, therefore, we kept our patient intubated to allow gradual weaning from mechanical ventilation. On next day, he was extubated in NICU and remained well. On 8th postoperative day, he started to develop respiratory distress and, on further work-up, was diagnosed with sepsis. He was continued on antibiotics, IV fluids and oxygen therapy for about a month. He gradually improved and finally was discharged home.

PATIENT'S CONSENT:
Informed consent was taken from patient's parent to publish this letter.

CONFLICT OF INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTIONS:
MY, AUH: Drafted the manuscript, done scientific literature search, and final approval of manuscript.

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Received: October 19, 2019; Revised: December 21, 2019; Accepted: December 24, 2019
DOI: https://doi.org/10.29271/jcpsp.2020.10.1115