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Case Report
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Emergency Surgery in a Child with Cretinism with Anticipated Difficult Airway Under Low-Dose Subarachnoid Block

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Abstract

A child with cretinism poses multiple anaesthetic challenges due to the associated mental and physical disability, deranged metabolic and physiologic functions, difficult airway and propensity to perioperative cardiorespiratory complications. Spinal anaesthesia in children is associated with remarkable cardiorespiratory stability and provides complete surgical anaesthesia. Here, we report a case that describes the first successful anaesthetic management of a child who was an unevaluated case of cretinism under subarachnoid block.

Keywords: Anaesthesia, cretinism, subarachnoid block

Introduction

Cretinism is a condition characterised by stunted physical and mental growth due to untreated congenital hypothyroidism with or without goitre (1-3). These patients pose many anaesthetic challenges due to associated physiological perturbations, difficult airway, mental retardation and short stature, especially for emergency surgeries (4). Currently, there is no report of anaesthetic management of a child with cretinism with anticipated difficult airway under sole regional anaesthesia. In the deficiency of any literature describing the anaesthetic management of these children, the general principles of managing overt hypothyroidism were kept in mind, anaesthetic implications were analysed and management strategy was formulated accordingly.

Case Presentation

A 10-year-old male child weighing 14 kg (height 97 cm) presented to the emergency room with an irreducible rectal prolapse. His medical history revealed complaints of frequent rectal prolapse in the last 1 year that had been treated with manual repositioning at home. There was a history of disproportionate growth, gross delay in developmental milestones, mental retardation and constipation. No clinical evaluation had been sought by his parents previously.

On physical examination, the child was of short stature, was able to follow verbal commands and had scaphocephaly, pectus carinatum, hypotonia, coarse facial features, hoarse cry, wide-open anterior fontanelle and macroglossia. There was no corneal clouding or hepatosplenomegaly. Heart rate was 70 beats min^{-1}, and rhythm was regular. There were bilateral conducted sounds in the chest. He could vocalise only monosyllables and could stand only with support. Airway was anticipated to be difficult in view of a large head, macroglossia, coarse facial features, short neck, irregular dentition and inability to cooperate.

On investigation, haemoglobin was 6.7 g dL^{-1}, urine examination was negative for sugar and albumin and kidney function test was normal. The patient was seen by a paediatrician in the preoperative holding area and was provi-

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sionally diagnosed with cretinism because of stunted growth, mental retardation, hypotonia, dull facial features, macroglossia, hoarse cry, history of constipation and wide-open anterior fontanelle. Blood sample was obtained for thyroid function test (TFT), and hydrocortisone 75 mg was administered intravenously (IV). In view of impending gangrene of prolapsed gut, the patient had to undergo an emergency rectopexy. Packed cell transfusion was started preoperatively. High-risk consent was obtained in view of anaemia, suspected cretinism, emergency nature of surgery with inadequate evaluation and optimisation and an anticipated difficult airway. After weighing the pros and cons, it was decided to perform surgery under low-dose spinal anaesthesia [1 mL bupivacaine, 0.5% hyperbaric and 0.4 mL fentanyl (100 µg mL^{-1} and 20 µg)]. A T10 level of sensory block was achieved. Intraoperatively, oxygen/nitrous oxide (50:50) was supplemented, and injection ketamine was administered twice in 5 mg increments for sedation. Blood loss was 80 mL. There was one episode of bradycardia (heart rate 52 beats min^{-1}) that responded to the administration of glycopyrrolate 0.15 mg IV. Injection emset 2 mg was administered. The rest of the intraoperative and postoperative periods were uneventful. The procedure lasted for 1.5 h. He received a total of 200 ml packed red blood cells and 300 ml Ringer's lactate. Postoperative analgesia was managed with regular scheduled diclofenac injections. The patient was planned for further investigations in the postoperative period. However, his parents absconded with the child before further evaluation could be done. The TFT report was later found to be consistent with severe hypothyroidism. Informed consent was obtained from the parents for publication of the clinical details of the child.

Discussion

Overt hypothyroidism leads to disturbed metabolic and physiologic processes in the body, thereby placing these patients at an increased surgical and anaesthetic risk (2, 4). Patients with hypothyroidism may have depressed myocardial function and cardiac output, reduced plasma volume, increased peripheral vascular resistance, hypoventilation, abnormal baroreceptor function, blunted ventilatory response to hypoxia and hypercarbia, sleep apnoea, anaemia, hypoglycaemia, hyponatraemia, hypothermia and impaired hepatic drug metabolism (2, 4, 5).

Main Points:

- Cretinism poses multiple anaesthetic challenges due to the presence of physiological derangements, difficult airway, short stature and mental retardation.
- General anaesthesia in the presence of overt hypothyroidism is associated with the risk of prolonged neuromuscular blockade and recovery along with the risk of hypothyroid coma postoperatively.
- Regional anaesthesia is an attractive choice in these children due to its lack of systemic depressant effects.
- Thyroid hormone replacement should be started preoperatively whenever feasible and steroid should be supplemented in severe cases.
- Subarachnoid block provides complete surgical anaesthesia with remarkable cardiorespiratory stability in children and should be considered for suitable surgeries in these patients.

One of the significant complications in these patients is increased risk of myxoedema coma postoperatively following general anaesthesia (GA). This condition has a mortality rate of 30%–60% even with optimal treatment (6). Therefore, elective procedures are considered contraindicated in patients with severe overt hypothyroidism and should be deferred until the patient has been rendered euthyroid (4, 5). In severe overt hypothyroidism, deranged physiologic processes, such as depressed myocardial function and hypoxic ventilatory drive, return to normal within 3–6 months on thyroid hormone replacement therapy, and the hypometabolic state becomes normal within 6 months (5, 7). A period of 2 weeks has been found to be adequate in normalising T4 levels. It is recommended that emergency surgery should be deferred for at least 48 h after the initiation of thyroid replacement therapy to allow for a latent onset of thyroid hormone activity (5, 7). In our case, the patient was posted for emergency surgery, and cretinism was suspected preoperatively but confirmed postoperatively; hence, thyroid hormones were not supplemented preoperatively. Concomitant adrenocortical suppression is thought to be present in severe cases, and steroid replacement is advised (8). Empirical hydrocortisone was administered perioperatively to the patient.

Preoperative supportive therapy should be initiated with IV fluids, blood transfusion, thermoregulation, correction of electrolytes and cardiorespiratory support. The risk of prolonged neuromuscular blockade and recovery from GA in hypothyroidism, along with the risk of hypothyroid coma postoperatively, prompted us to select subarachnoid block for this child.

Spinal anaesthesia is an uncomplicated and effective technique that provides a rapid onset and profound analgesia, sympathetic and motor block in the lower part of the body. It provides effective surgical anaesthesia with minimal physiological changes or adverse effects (9). This advantage is important in high-risk children where it is desirable to avoid tracheal intubation and mechanical ventilation. The risk of potential difficult airway, opioid-related respiratory depression and sensitivity to muscle relaxants is more pronounced in cretinism (1, 2, 4). Low-dose spinal anaesthesia avoided any physiological derangements related to polypharmacy and postoperative effects of anaesthetics and muscle relaxants. The recommended dose for children weighing 5–15 kg is 0.4 mg kg^{-1} (0.08 mL kg^{-1}) bupivacaine (9). For this child, the dose

was 1.12 mL, and he was administered 1 mL bupivacaine intrathecally. The duration of spinal anaesthesia in children is shorter due to their highly vascular pia mater leading to faster reabsorption of the injected drug. Hence, fentanyl was added as an adjuvant to improve the duration of block and prolong the postoperative analgesia. The patient was very comfortable postoperatively and could be managed with nonsteroidal anti-inflammatory drugs only.

Effective early postoperative analgesia also created the ideal physiological conditions for the recovery period in this child. An awake and painless child is easily manageable postoperatively. Additionally, efficient pain control reduces adverse effects due to the neuroendocrine stress response.

Of great advantage to children is the fact that spinal anaesthesia is characterised by remarkable haemodynamic stability (9). Additionally, regional anaesthesia can significantly attenuate the surgical stress response. Wolf et al. (10) reported that in infants undergoing major surgery, central neuraxial block was more effective than high-dose opioids in suppressing cardiovascular and stress responses. Severe neuroendocrine stress response may increase postoperative morbidity and mortality.

Conclusion

Cretinism presents as an anaesthetic challenge owing to multisystem involvement. Regional anaesthesia is a safe option in these patients and should be considered when feasible.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this case.

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