Anaesthetic considerations in a child with rickets and craniosynostosis for linear strip craniectomy and frontal advancement

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Sir,

Craniosynostosis is the result of premature closure of the cranial vault sutures.\(^1\),\(^2\) Besides the functional impairment, there are often problems concerning the social integration of patients caused by the grotesque skull deformities.\(^3\) Craniosynostosis can be nonsyndromic or syndromic (e.g. Apert’s syndrome and Crouzon’s syndrome).\(^2\) Craniosynostosis has been observed in association with a number of maternal metabolic disorders including hyperthyroidism, rickets, Hurler syndrome, Morquio syndrome, beta-glucuronidase deficiency, mucolipidosis III, and a host of haematological disorders.\(^2\)-\(^4\) We report the anaesthetic considerations of a child of rickets and craniosynostosis planned for linear strip craniectomy and frontal advancement.

An 11-month-old male child weighing 6 kg presented in the neurosurgical clinics with chief complaints of progressively increasing bilateral proptosis since birth and abnormal shape of head since 3 months. He had a history of decreased sleep, increased sweating and increased frequency of micturition. Wrist X-ray showed widening of distal end of ulna, suggestive of rickets. He was administered vitamin D megadose. He continued to have persistently low calcium levels and phosphate levels.

The child was diagnosed with bilateral coronal craniosynostosis and it was planned to perform linear strip craniectomy and frontal advancement on the child. The respiratory and cardiovascular examination revealed no abnormality. On investigation, the haemoglobin was 7.2 g/dL and the child received two paediatric units of packed red cells transfusion and his haemoglobin improved to 11.2 g/dL. His serum calcium level was 8.9 mg/dL, phosphate level was 5.5 mg/dL and alkaline phosphate level was 980 IU.

In the operation room, routine monitors were attached. Anaesthesia was induced with 8% sevoflurane in oxygen and thereafter 22G intravenous access was secured. Then, 20 µg fentanyl and 20 mg rocuronium were administered intravenously. Airway was secured with size 4 mm ID endotracheal tube. Capnography and temperature monitoring were initiated Anaesthesia was maintained with 1% isoflurane in oxygen and nitrous oxide (50:50). The child was positioned for surgery. Top ups of fentanyl (total 30 µg) and rocuronium were administered, as guided by neuromuscular monitor. Intraoperatively, 50 mL of 20% mannitol was administered. Blood loss was 90 mL and was replaced with crystalloids adequately. The surgical duration was 4 hours. At the end of surgery, residual neuromuscular blockade was reversed and trachea extubated. The child had an uneventful recovery.

Our case of craniosynostosis was probably secondary to hypophosphataemic rickets. In our case, apart from cosmetic reason, considerations of the progressively increasing vision-threatening proptosis and risk of neurological impairment required urgent surgical repair.\(^1\) The perioperative concerns in our patient included risk of corneal ulceration, airway related, rickets, air embolism, blood loss, and prolonged surgery with risk of head and neck oedema.

These patients should have had a complete multidisciplinary evaluation to rule out any syndromic association. Special attention should be directed to signs of increased intracranial pressure.\(^3\) The poorly protected eye is exposed to the risk of corneal ulceration. The airway securing is difficult because of not only being an infant but also having abnormal facial features. The surgery on the head also had its implications for accidental extubation. The decreased level of calcium was a concern in view of neuromuscular function and risk of fractures during positioning. Venous air embolism has been a reported complication of craniosynostosis repair.\(^1\),\(^2\) The inevitable blood loss is another concern in these procedures. Adequate venous access is critical. Embarrassment of venous drainage and lymphatics leads to significant oedema around the cranium in the first 24 hours. Generalised oedema can be minimised by optimising crystalloid and blood administration during the case.

The perioperative management of children who have these congenital malformations requires multidisciplinary care.
Brief Communications

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Sir,

Central venous pressure (CVP) monitoring is a simple, relatively inexpensive method of assessing a patient's circulating blood volume, cardiac status and vasomotor tone. It is essential to be aware of the inherent fallacies and inadequacies of the information derived. Inaccurate measurements are often obtained by the aberrant lodgement of the central venous catheter (CVC) tip.

In continuation of the previously published letter to editor concerning misdirected CVC, we describe an unusual case of CVC coiling in the superior vena cava (SVC) leading to falsely high CVP measurement. A 55 year old male patient was brought to the emergency room (ER) with head injury, blunt trauma abdomen with haemodynamic instability. He was further posted for an emergency laparotomy. In view of the clinical condition of the patient and the need to know intravascular volume status, a 7 F rench triple lumen CVC was inserted in the right internal jugular vein (IJV) in the operating room (OR). All the three ports were checked for free flow of blood and the CVC was fixed at 11cm at skin level. On connecting the transducer to the monitor, ideal waveform was absent. Intra operatively CVP tracing was suboptimal despite the change of transducer, the cable, flushing the unit and repeated zeroing. Post operatively the patient was shifted to intensive care unit on ventilator support for further management. Chest radiograph revealed coiled CVC in the SVC [Figure 1]. Hence it was removed and right subclavian vein was cannulated.

The correct placement of the CVC tip is an important factor in obtaining accurate CVP measurements. Malposition of a CVC may occur at the time of insertion or later as a result of spontaneous migration due to anatomic positioning or pressure changes within the thoracic cavity.

There has been case report on CVC folding back during guide-wire removal inside IJV. In our case CVC coiling inside SVC was unusual as it is a large calibre vessel with high flows. There was no anatomical vascular abnormality and no manufacturing defect in CVC. The abutting of the guide wire against the wall of the SVC probably caused the coiling of CVC in the SVC. The J tip of guide wire probably was unknowingly directed cephalad while insertion, which could have caused CVC to further angulate in the upward direction over the guide wire.

In conclusion, inaccurate CVP measurements or inability to obtain an ideal wave from tracing are suggestive of an undesirable location of the catheter tip. Awareness of this possibility and careful review of the CVC tip position on X-ray pictures in suspicious cases are important. Inaccurate CVP readings lead to improper assessment of the intravascular status of the patient. Careful clinical co-relation under such circumstances is essential. Roentgenograms after insertion of CVC are essential to eliminate this problem, which is often encountered in clinical practice.