

Letters to Editor

Bilateral ESP block: Savior in patient with Crouzon syndrome

Dear Editor,

Crouzon syndrome is a rare genetic disorder (incidence 1.6 in 100,000 patients), characterized by craniofacial dysostosis caused by premature fusion of cranial sutures due to mutation in one of the FCFR (FGRR2) genes.^[1] This affects the growth of skull and head resulting in abnormal shape and development of head and face, manifested as brachycephaly, shallow orbits, protruded eyeballs, prominent foreheads, midface hypoplasia, hypoplastic jaw, relative mandibular prognathism, high-arched palate with crowded teeth, and sometimes spine abnormalities.^[2] Here, we are presenting a

successfully managed case of Crouzon syndrome, posted for total abdominal hysterectomy (TAH).

A 45-year-old female with Crouzon syndrome, with loss of vision and exposure keratitis due to severe proptosis, brachycephaly, neck movement limitation, maloccluded multiple loose and missing teeth with moderate kyphoscoliosis was posted for TAH for cervical fibroid [Figure 1]. On further evaluation, we found that there was associated pulmonary hypertension (pulmonary artery pressure = 38 mm Hg) and hypothyroidism for which she was on medication for 5 years. Her laboratory parameters were normal except low hemoglobin (7.7 g/dL) despite two units of cross matched packed cell transfusion, most probably due to menorrhagia. Her intelligence and mentation were not affected. Our main concern in this case was difficult airway in view of limited neck movement, abnormalities in shape of head, face and



Figure 1: Proptosis, brachycephaly and maloccluded loose and missing teeth in Crouzon syndrome

dental anomalies, associated pulmonary hypertension, and moderate kyphoscoliosis imposing procedural difficulty for central neuraxial block. Administering neuraxial blocks can be technically difficult in patients with Crouzon syndrome due to vertebral fusion and presence of scoliosis or kyphosis.^[3] There was also high probability of eye injury due to severe proptosis while mask ventilation and intubation. Thus, we planned for monitored anesthesia care with bilateral ultrasound-guided erector spinae plane (US-ESP) block with dexmedetomidine sedation. Patient was educated about ESP block and numeric rating scale (NRS) for pain assessment. In the operation theater, noninvasive blood pressure, SpO₂, and electrocardiogram monitoring were started. After positioning the patient in sitting position, bilateral US-ESP block was given with 20 mL of 0.25% levobupivacaine, at the level of second lumbar vertebra and catheter was secured in that myofascial plane for perioperative continuous infusion of 0.125% levobupivacaine at the rate of 6 mL/h. Intravenous bolus of 50 µg dexmedetomidine was given over 10 minutes followed by infusion at the rate of 5 µg/h to achieve Ramsay sedation score of 3. Block level till sixth thoracic vertebra was assured before the commencement of surgery. Oxygen supplementation was done through nasal cannula at the rate of 3 mL/min. Intraoperative period was uneventful. Patient was followed for two postoperative days and was comfortable and stable. NRS was always less than 4.

US-ESP block is a myofascial plane block, described by Forero *et al.*^[4] where drug diffuses in a plane below erector spinae muscle and provides analgesia by blocking dorsal rami, ventral rami as well as rami communicantes of spinal nerves. The ESP block is easy to administer, safe, with no hemodynamic changes and good analgesia that can be used for TAH as an alternative technique to general and regional anesthesia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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