Pectus carinatum repair in an adolescent with hyperhomocysteinaemia: Anaesthetic implications

Sir,

Pectus carinatum is a chest wall deformity that occurs less frequently and surgical correction for this chest wall deformity is mostly for cosmetic reasons. Anaesthesia for this procedure may be complicated by associated systemic disorders such as Marfan's disease, homocystinuria, Prune belly syndrome, Morquio syndrome, osteogenesis imperfecta, Noonan syndrome, mitral valve prolapse, scoliosis or other collagen vascular diseases.^[1] Although the surgical aspects of pectus carinatum repair are extensively reported, literature on anaesthetic management is sparse. We report the successful management of a case of pectus carinatum repair in an adolescent patient with hyperhomocysteinaemia and kyphoscoliosis.

A 14-year-old boy weighing 40 kg presented to the hospital with complaints of protuberance of the lower chest since birth which had significantly increased in the last 2 years. Computed tomography of the chest revealed chondrogladiolar type of pectus carinatum with Haller index of 1.6 (Haller's index, the ratio of transverse diameter of thorax to antero-posterior diameter at lower third of sternum, normal value = 2.5) and thoracic kyphoscoliosis to left [Figure 1]. Pulmonary function testing showed moderate restrictive pattern. A year earlier, he had suffered from the left upper motor neurone Bell's palsy, which was self-limiting. Evaluation for early onset of cerebrovascular disease revealed homocysteinaemia with homocysteine levels of 18.5 µmol/l. He was treated with folic acid, Vitamin B6 and B12 supplements. The patient now desired to have a repair of pectus carinatum which was scheduled under general anaesthesia. Pre-operatively, vitamin supplements were continued.

In the operation theatre, intravenous (iv) line was inserted and American Society of Anesthesiologists standard monitoring was connected. An epidural catheter was placed in T7–8 space for intra and postoperative analgesia. Epidural analgesia was initiated with a bolus of 7 ml of 0.25% bupivacaine followed by an infusion of 0.125% bupivacaine at 8 ml/h. Elastic stocking was used in the legs to avoid venous stasis. The patient was pre-medicated with fentanyl 100 μ g, midazolam 1 mg, induced with thiopentone 200 mg and tracheal intubation was

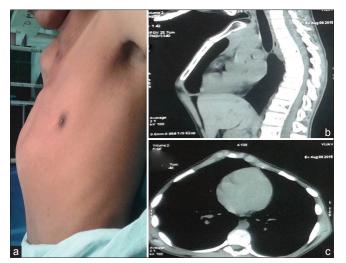


Figure 1: (a) Image of the patient chest showing pectus carinatum. (b and c) computed tomography scan chest of the patient (sagital and transverse sections, respectively) showing pectus carinatum

facilitated with rocuronium 1 mg/kg. The patient was intubated with a 7-mm-cuffed endotracheal tube and ventilated in volume control mode with a tidal volume of 300 ml. The respiratory rate was adjusted to EtCO₂ of 35–38 mm Hg. Anaesthesia was maintained on oxygen in 50% air, sevoflurane and infusion of atracurium. Bilateral pectoralis major flaps were raised and outgrown portions of third to seventh costal cartilages were resected [Figure 2]. Costochondral junctions were fixed with miniplates and screws. Finally, it was covered with the pectoralis major flaps [Figure 2]. The total surgical duration was 7 h. Continuous temperature monitoring and random blood sugar monitoring were done. Forced air warming was used to maintain normothermia. In view of extensive dissection of thoracic wall, the patient was electively ventilated post-operatively and extubated after 6 h. In the post-operative period, adequate analgesia and early mobilisation were stressed upon to reduce the risk of deep vein thrombosis (DVT).

Pectus carinatum is chest wall deformity characterised by anterior protrusion of sternum due to overgrowth of costal cartilage and mainly affects males.^[1,2] During growth spurt of puberty, the condition becomes more obvious and consequently, changes in behaviour and personality may frequently occur. Pectus carinatum may be associated with Marfan's disease, homocysteinuria, Prune belly syndrome, Morquio syndrome, osteogenesis imperfecta, Noonan syndrome and mitral valve prolapse.^[1]

The most common treatment is use of sternal braces. Surgery may be opted in patients not responding to

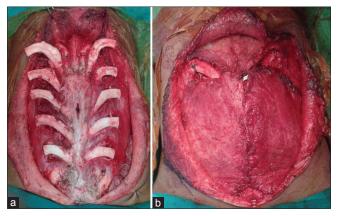


Figure 2: Intra-operative images showing repair of pectus carinatum. (a) The exposed costochondral joints of $3-7^{th}$ ribs. (b) Bilateral pectoralis major flaps covering the repaired costochondral joints

braces, older children with hard sternum, in patients with pain or for cosmesis. Surgical repair of pectus carinatum has also shown to improve mental health and quality of life.^[3,4] The surgical procedure involves resection of overgrown costal cartilages, osteotomy of sternum and fixation with metal plate and screws. In our patient, additional pectoralis major flap was taken to cover the implants and thus avoid infection.

Hyperhomocysteinaemia is a genetic disease leading to accumulation of homocysteine causing multisystem manifestations. Normal levels are within 7-10 µmol/l. Levels between 15 and 30 µmol/l are considered mild to moderate.^[5] Homocysteinaemia causes oxidative vascular endothelial damage and creates procoagulant state by activating platelet aggregation and adhesion.^[6] Thus, it can increase the risk of DVT in prolonged surgeries. Hence, we used elastic stockings in the intra-operative period. In patients with homocysteinaemia, increased levels of methionine leading to increased insulin release can cause hypoglycaemia. Thus, we monitored random blood sugar levels intra-operatively. Nitrous oxide was avoided as it may increase homocysteine levels by inhibiting methionine synthase.^[7]

Pulmonary function restriction in our case could be partly due to pectus and partly due to kyphoscoliosis. Surgery does not improve pulmonary function in pectus carinatum unlike pectus excavatum.^[8] Thus, pre-existing restrictive ventilatory function may be further compromised by pain and optimising post-operative analgesia is the major concern in these surgeries. Continuous epidural infusion is most commonly used for providing analgesia in pectus correction surgeries, but patient controlled iv analgesia has been found to be equally effective.^[9,10] We could place the epidural catheter without much difficulty because of less severe spine deformity in our patient and achieved satisfactory analgesia.

The associated comorbidities, pain and respiratory dysfunction may complicate the benign cosmetic procedures for pectus carinatum. Attention to DVT prevention, adequate analgesia and early ambulation aid in successful management in these patients.

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Conflicts of interest

There are no conflicts of interest.

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