



PEDIATRIC SURGERY *Update**

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Pectus Arcuatum

Anterior chest wall deformities are not rare. They consist of pectus excavatum as the most frequent form of chest deformities, and by protrusion deformities such as pectus arcuatum and carinatum. Pectus arcuatum, also known as pouter pigeon chest, Currarino-Silverman syndrome, chondro-manubrial deformity or type two pectus carinatum, is a rare and complex congenital chest wall deformity whose main feature is protrusion and early ossification of the sternal angle of Lewis associated with bilateral deformity of the 2nd to 4th cartilages. It may also be associated with a depressed lower sternum. It also involves a wavelike deformity, a mixed form of excavatum and carinatum features, either along a longitudinal or along a transverse axis. The visual appearance of pectus arcuatum is formed by the costal cartilage protrusion. In most cases pectus arcuatum is a cosmetic defect. Though the diagnosis is established by physical exam, chest films and CT-Scan with 3-D reconstruction are needed if repair of the sternal defect is warranted. Imaging using thoracic CT scans and customized aided design virtual simulation allows the surgeon to predetermine the specific cutting angle for each patient and therefore design a cutting template tailored to the individual deformity. The surgical repair of this rare deformity requires a modified open Ravitch technique. Patients are usually young adults without comorbidities and no special preparation is needed. The basic steps in the surgical correction described by Ravitch consist of bilateral parasternal and subperichondrial resection of the deformed costal cartilages, detachment of the xiphoid process, transverse wedge osteotomy at the upper edge of the sternal depression, and bending of the sternum to straighten its course, securing the corrected position of the sternum. Satisfactory overall results occur in 98% of patients. The Ravitch technique has a risk of growth limitation to the thoracic cage due to a wide resection of the rib cartilages, the reason that the repair is not undertaken until the child has acquired a rigid skeletal structure later in life. Pectus arcuatum can be successfully corrected by Ravitch-type of chondrosternoplasty. Due to necessity to resect cartilages, late puberty or adulthood is preferred, since by that age the growth of ribs have finished. Repair of pectus deformity in children that might need future cardiac surgery has revealed that concomitant surgery is contraindicated before adolescence because pectus deformities may spontaneously disappear or recur after early sternal surgery. Congenital heart defects are reported occasionally as well as simultaneous Poland syndrome. Concomitant surgery of cardiac defects and pectus deformity is a reliable strategy in adolescent and adults offering long-term results. The modified Ravitch technique is more adequate as it can be used in all types of deformities and in concomitant surgery allowing optimal operative exposure during cardiac procedures, easy postoperative reentry and resuscitation maneuvers if needed.

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Imposter Syndrome

Imposter syndrome refers to a feeling of self-doubt or innate fear of being discovered as a fraud or non-deserving professional, despite their demonstrated talent and achievements. Imposter syndrome is characterized by a chronic sense of self-doubt coupled with a constant worry of being discovered as a fraud. Imposter syndrome is more prevalent in high achievers, women, and under-represented racial, ethnic, and religious minorities. Impostor syndrome is increasingly recognized as a condition between physicians and physicians in training. Despite remarkable academic and professional achievements affected individuals' beliefs that they were unintelligent. For affected individuals, imposter syndrome can lead to burnout, psychological distress, emotional suffering, and serious mental health disorders, including chronic dysphoric stress, anxiety, depression, drug abuse and suicide. Most cases start early during high school or college. The true incidence of imposter syndrome is unknown in medical professionals. In the US, among medical students the rate of imposter syndrome was 49% in women and 24% in men, and among residents the rate was similar. The root cause of imposter syndrome is not known, but it has been related to depression and anxiety, which are both present among residents, with suicide being the second most common cause of death among residents. Imposter syndrome among general surgery residents is not only prevalent but severe with 76% of residents reporting either significant or severe imposter syndrome. Neither sex nor age correlates with the presence of, or level of, imposter syndrome in the general surgery resident population. It is believed that imposter syndrome is an incidentally protective mechanism encouraged by the hierarchical culture of surgical training by which residents are encouraged to self-regulate their decision-making process. By constantly downplaying their own accomplishments, those suffering from imposter syndrome may sabotage their own career. Institutions must address imposter syndrome by increasing the visibility of the problem, providing access to mental health coaching, and establishing supportive organization policies. Institutions and residency programs should provide training for mentors to help them recognize the negative consequence of the imposter syndrome. Medical educators must recognize that it is not just the underperforming or failing learners who struggle and require support, and medical culture must create space for physicians to share their struggles. The Accreditation Council for Graduate Medical Education (ACGME) requires residency programs to support residents' well-being via established policies and programs. Imposter syndrome has been linked to burnout and suicide in residents and understanding how to combat it may help

improve resiliency in residents. Imposter syndrome has been linked to resident burnout and discussing imposter syndrome is viewed as an effective intervention to promote resident wellness and resiliency. When creating wellness interventions, residency programs should consider addressing imposter syndrome.

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Short Bowel Syndrome

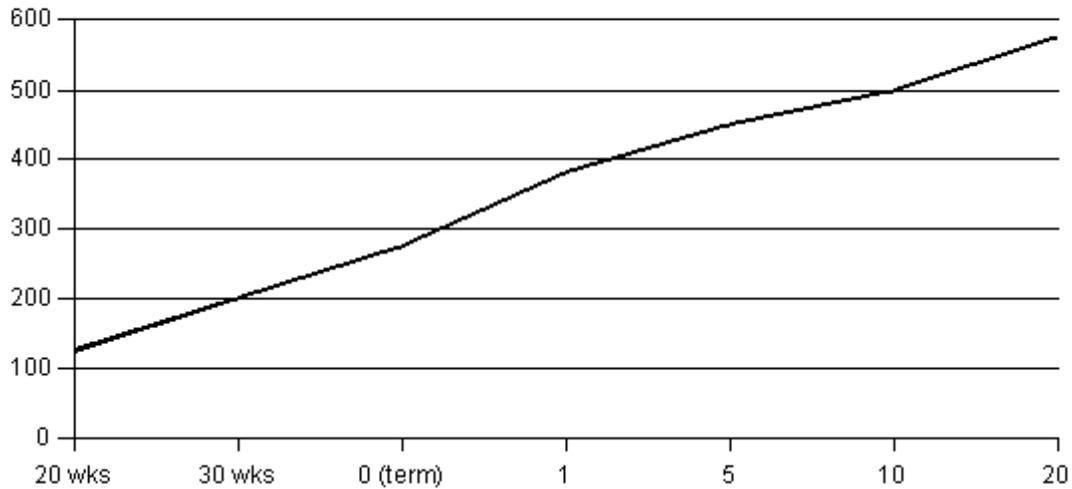
Short bowel syndrome (SBS) refers to a compromised bowel absorptive capacity due to severely reduced mucosal surface resulting in diarrhea, water-electrolytes imbalances, and protein malnutrition. SBS is the most common cause of intestinal failure in children. Most underlying conditions that lead to major loss of intestine in neonates have their origin in intrauterine life. SBS usually occurs after extensive bowel resection, either congenital or acquired, such as that associated with small bowel atresia, complex gastroschisis, midgut volvulus and necrotizing enterocolitis. The most common acquired cause of SBS is NEC with 30% in most reported series. Factors influencing outcome in SBS include underlying diagnosis, type of segments preserved, stoma vs primary anastomosis, presence of ileocecal valve and the age of the child at the time of surgery. Massive resection stimulates modification in thickness and length of the muscle layer and villi crypts. Distension of the remaining bowel is the most common consequence after massive resection. Massive jejunal resections are better tolerated than significant ileal resections. Ileal resections are associated with impaired resorption of Vitamin 12, bile salts and fatty acids. Three anatomical subtypes of SBS: (1) small bowel resection with anastomosis and intact colon; (2) small bowel resection with partial colon resection; (3) small bowel resection with high output jejunostomy. Type 1 has the best potential for adaptation, while type 3 the least. With the advent of parenteral nutrition support survival of SBS improved significantly. Management of SBS aims to promote adaptation of the remnant bowel. Parenteral nutrition (PN) provides nutrition while the bowel achieves intestinal autonomy. The bowel should be use for feeding as much and early as possible to stimulate adaptation. Oral feeding maintains sucking and swallowing functions, promotes release of epidermal growth factor from salivary glands and increases GI secretion of trophic factors. Breast feeding should be encouraged. Long term PN leads to sepsis, cholestasis due to liver failure and death. Key predictors of mortality in SBS include cholestasis (conjugated bilirubin > 2.5 mg%) and percentage of small bowel length. A small bowel length greater than 10% of expected for a given gestational age is highly predictive of survival (see normal bowel length in accordance with gestational age graph). Presence of an ileocecal valve and percentage of

small bowel length are primary predictors of weaning PN. Surgical approaches to maximize bowel digestive and absorptive function are important in the management of SBS. These include stoma closure, bowel continuity restoration, resection of strictures and closure of fistula. When the bowel is short, dilated, and static children might benefit from longitudinal intestinal lengthening and tapering (Bianchi) or serial transverse enteroplasty (STEP) procedures. The UGIS provide accurate estimates of bowel diameter and length use to operative planning. Surgical bowel lengthening should be considered in any chronically PN-dependent child when there is substantial bowel dilatation and symptoms of small intestinal bacterial overgrowth regardless of remaining bowel length. Medical approach to SBS include antidiarrhea/ antimotility agents and controlling acid/base balance. Hypersecretion of gastrin and gastric acid occurs in children after extensive small bowel resection which should be managed. Octreotide inhibits gastrin and diarrhea prolonging transit time. Intestinal bacterial overgrowth frequently seen in SBS is managed with probiotic and antibiotic therapy. Promising hormonal therapy include glucagon-like peptide 2 hormone (Teduglutide) produced by the L-cells of the terminal ileum. Teduglutide has a trophic effect on the bowel, promotes absorption and adaptation. The future of SBS might lie in an artificial grown and engineered harvested intestine. Micronutrient deficiencies are frequent during intestinal rehabilitation for SBS. The most common micronutrient deficiency include zinc, copper, vitamin D and phosphorus after the transition to enteral nutrition. With liver failure and reduced venous access, bowel transplantation becomes the treatment of choice.

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Bowel Length in cm



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