

PEDIATRIC SURGERY Update* Vol. 59 No. 02 AUGUST 2022

Perianal Crohn's Disease

Crohn's disease is a transmural bowel inflammatory disease affecting any age, involving all segments of the intestine including extraintestinal sites. Almost one-fourth of Crohn's cases occur in children younger than 18 years of age. Perianal Crohn's disease (PCD) accounts for 10-40% of children with Crohn's disease. PCD has a wide variety of clinical manifestations such as skin lesions, anal canal lesions, anoperineal abscesses or fistula, anovaginal fistulas and cancer (adenocarcinoma and squamous cell carcinoma). Skin lesions can be further subdivided into tags, hypertrophic papilla, or hemorrhoids, while the canal lesions include fissures, ulcers, or stricture/stenosis. Fissures and skin tags are the most common presentation of pediatric PCD. The symptoms of PCD include pain, itching, bleeding, purulent discharge, and incontinence of stools. Initial inflammation in the affected rectum forms ulcers or shallow fistulas, they subsequently extend into the deep or penetrating fistulas with persistent exposure to feces and pressure caused by defecation; infected anal glands penetrate the intrasphincteric space and progress to form fistulas or abscess. Manifestations of PCD have three categories: 1) tissue destruction (anal fissures, tags, deep ulcers lined with granulation tissue), 2) fistulas and abscess (multiple, complex, frequently related), 3) rectal stricture due to long standing disease. Spectrum of PCD can be mild or severe leading to constipation, fecal incontinence, recurrent infections, sepsis, compromise sexual function, diminished quality of life, which could end in diverting colostomy or ileostomy. PCD is usually painless and is diagnosed by physical exam. Endoanal US and MRI can further aid in diagnosis, rectoanal involvement, involvement of other adjacent structures and management. Most useful radiological study to evaluate abscess and fistulas is MRI. Evaluation of PCD starts with external/rectal examination followed by MRI, and exam under anesthesia performed by experienced surgeon. Presence and extent of intestinal disease can be further determined using a variety of endoscopic, anoscopy, ultrasound, and imaging tests (CT, MRI, fistulography). Skin tags do not resolve completely with treatment but remain present and benign. Fissures will often heal completely with minor nitrate-based therapy. Is clinically relevant to use Bell's classification of fistulas into simple and complex. Simple fistulas involve a low intersphincteric or trans-sphincteric location, single short tract, internal opening lower and closer to anal verge with the external opening near the anal verge without abscess. Complex fistulas involve sphincter muscle/anorectal ring, are multiple, branch with or without abscess, with internal opening of the fistulous tract above sphincter muscles and external opening further away from anal opening. Simple fistulizing disease if superficial and confined to the anal canal healing spontaneously in 50% of cases. Rectovaginal and complex fistulas (high intersphincteric, high trans-sphincteric, extrasphincteric or suprasphincteric) rarely heal without therapy. Rectal strictures predict poor outcome, diagnosis is often delayed, most are in the distal rectum associated with proctitis, with most children responding to anal dilatation, preferably under anesthesia. Tight obstructive stricture should be considered for ostomy diversion. Medical management of PCD include local measures such as warm sitz baths, bowel regulation with fiber products, or antidiarrheal medication. Steroids should not be used during active disease. Antibiotics, immunomodulator and biologic agents are very beneficial to these patients. Antibiotics are first line therapy reducing fistula associated pain and drainage include metronidazole or ciprofloxacin, though symptoms usually recur immediately after antibiotic discontinuation. Immunomodulation using azathioprine or 6-mercaptopurine is effective, but slow and incomplete: leucocyte count and liver transaminase levels should be monitored. Biologic agents are the mainstay therapy for complex PCD. Biologics refer to anti-tumor necrosis factor antibodies which are potent anti-inflammatory agents such as infliximab and adalimumab. Anti-TNF biologic therapy using adalimumab without surgery is effective therapy for healing fistula in children with moderate to severe PCD in approximately half of the patients. The success of surgery management during acute tissue inflammation is limited, and fraught with complications. Limitation includes draining abscess and relieving acute symptoms. Perineal abscess cavities should be drained. Drain both internally and externally, keep communication between then with silk or silastic. Seton placement, which is reserved for the management of complex fistulas, can be left in place indefinitely, they usually deteriorate and fall in around one year, cause no harm and is well tolerated. Once inflammation subsides considerably, elective surgery to deal with fistula and stricture should followed. Combined surgical and biologic therapy produce better healing of perineal disease, longer effect duration and lower recurrence rate compared with either treatment alone. Stubborn fistulas that persist after medical therapy can be managed with fibrin plug or fibrin glue. Severe PCD can be managed with colostomy or ileostomy with a high risk of becoming permanents. With minimal colitis a sigmoid diversion is performed, with colonic involvement and ileostomy is utilized.

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3D Printing

3D printing (3DP), also known as additive manufacturing, is the construction of a threedimensional object from either a computer-aided designed or a digital 3D model. The final product is deposited, joined or solidified under computer control with materials being added together layer by layer to create a synthetic part. The 3DP process typically consists of depositing material layer-by-layer in a semiliquid, liquid or powder form and solidifying using light energy, an electron beam, chemical binders, or heat and allowing to solidify at room temperature. 3DP modeling can process a wide array of materials such as metals, polymers, or even ceramics into the desired shape. The technical pathway to create a physical 3D anatomical model starts with image acquisition, postprocessing segmentation, preprint preparation and the printing process itself. The key advantage of 3D printing in medicine is the ability to produce very complex shapes or geometrics that would be otherwise almost impossible to construct by hand including hollow parts or parts with internal truss structures such as body cavities. 3D printing technology can model a specific patient anatomy based on CT/MRI imaging using specific software for such purposes. The created model can be a complex congenital malformation in any part of the body. Threedimensional visualization system provides surgeons with more in-depth knowledge of the topographic anatomy. This construction provides the potential for teaching and training after successfully producing accurate models of complex or rare anatomy.3DP technology is rapidly growing in many surgical fields such as maxillofacial reconstruction, craniofacial, ENT, urology, orthopedics, cardiac, cancer, hepatobiliary, plastic, and pediatric surgery. The 3D printed models created are highly fidelity to the original organs and capable of demonstrating spatial relationship of target area of organs involved in disease or malformations.3D printing can help better understand crucial surgical anatomy of solid organs not easily accessible through conventional imaging. Vivid demonstration of spatial relationship of three-dimensional objects is of great value in understanding complex surgical anatomy and subsequent operation planning. The use of 3D printing technology enables the creation of simulation models for teaching and practicing purposes such as learning to do laparoscopic or thoracoscopic procedures such as pyloromyotomy and repair of esophageal atresia respectively. Congenital heart diseases such as atrial septal defects, ventricular septal defects, Fallot tetralogy and transposition of the great vessels have benefited from 3DP modeling technology. Interventional cardiology has successfully used 3DP model to stimulate surgical stent placement and assessment of flow. In neurosurgery, surgeons use the 3DP models for comprehending the spatial relationship of brain lesions to the neural pathways, blood vessels, and functional anatomy. Orthopedic application of 3DP with MRI data is mainly used in resection planning for oncological tumors such as osteosarcomas and osteochondromas. Surgeons have reported more precise resections of bone tumors, with less blood loss, shorter operative time, and reduced radiation exposure during surgery. Urological application of 3DP includes kidney tumors resection planning and increase ability to plan for nephron-sparing procedures. Comparison between 3DP models of normal and abnormal anatomy is utilized to educate trainees and preoperative counseling of patient. Hence 3DP clinical applications include teaching, developing a diagnosis or plan (intervention and simulation), procedure performance using the models, and materials (shaping devices to mold growing anatomy or substitution prosthetics). Costs are still high.

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Sea Urchin Injuries

Saltwater aquatic animals can cause significant injury and morbidity to humans, specially to children. Aquatic ecosystems are usually visited for commercial (fishing) and recreational (bathing and aquatic sports) purposes. Trauma and envenomation can be caused by sea urchin spines. Sea urchins are invertebrates and member of the echinoderm's family with radial symmetry. There are over 700 known species of sea urchins with approximately 80 species toxic to humans. As slow moving non-aggressive and bottom dwellers sea animals they are found in deep water, rocky inclines, and coral reefs. Spines of calcium carbonate and pedicellaria are attached to the tegument. The spines can be hollow or solid with toxins associated with them or their thin epithelial layer. The spines can be short and thick, or long and fine penetrating skin through footwear or bathing suit. The toxin substance identified in the pedicellaria, or spines include steroids, serotonin, glycosides, cholinergic substances, histamine, and bradykinin. Primary injury of sea urchin is through spine penetration with the most common affected areas being feet (80%), ankles and hands (15%). Diagnosis is usually based on clinical findings and history. Injuries by urchin spines are usually seen in divers, snorkelers, bathers, and fishermen. Reactions related to sea urchin can be classified as immediate and delayed. Spine penetration causes immediate intense pain, bleeding, erythema, edema, and local myalgia. Should a joint be penetrated synovitis may occur. With more than fifteen spines injury systemic symptoms can occur such as paresthesia, radiated pain, hypotension, muscular weakness, dyspnea, aphonia, deafness and even death. Spine fragment retention can cause further infection, granulomas and foreign body reaction with pain and edema lasting for weeks with vesicular reaction due to delayed hypersensitivity. Sea urchin granulomas are believed to be delayed reaction as a result of the spine remnant in the wound. The granulomas develop due to foreign body reaction against inorganic substances in the spine. Clinically it manifests as a firm, painless, nonsuppurative nodule with a central depression and hyperkeratotic surface, especially if localized in the hand. They are pathologically described as sarcoid type. Tenosynovitis can occur if joints or tendons sheaths are penetrated by the spine. Impairment of joint function and severe destructive effects with functional loss can result when the inflammation takes the form of granulomatous synovitis or chronic chemical synovitis. The spine fragments may or may not be seen in simple x-ray films. Ultrasound or MRI imaging can be helpful in doubtful cases. Treatment of sea urchin spine injury in most of the cases is conservative. The management of sea urchin injuries is immediate immersion of the affected area in warm water (110-115 F) for 30-90 minutes. This makes the toxin and proteinaceous irritants inactive relieving pain. Some sources recommend mixing water with vinegar in a 1:1 ratio. Spines and pedicellaria should be removed by irrigation with water and soft traction. Observation and supportive treatment for allergic reactions, muscular paralysis and respiratory distress is recommended with more than 15 spine injuries at a time. Surgical exploration is indicated when fragments are affecting joints, tendons, or neurovascular structures to prevent delayed complications. Local or systemic antibiotics is recommended for secondary infections. Difficult to extract spines can be destroy using erbium:YAG laser ablation. Laser utilization is preferred in areas such as the heel of the foot.

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