



PEDIATRIC SURGERY Update*

Vol. 53 No. 06 DECEMBER 2019

Choledocholithiasis in Sickle Cell Disease

Sickle cell disease (SCD) is a vaso-occlusive hemolytic anemia that causes pigmented gallbladder and common bile duct stones (choledocholithiasis) due to increase red blood cell destruction. Elevated serum bilirubin is produced from the breakdown of heme release from hemolyzed erythrocytes. Choledocholithiasis can occur concomitantly with gallstones (secondary) or years after removal of the gallbladder (primary). Most cases (95%) of bile ducts stones in SCD are from migration of gallstones from the gallbladder. Diagnosis of primary common bile duct stones depends on the following criteria: 1) previous removal of the gallbladder with or without bile duct exploration, 2) at least two-year asymptomatic period after surgery and 3) no evidence of biliary stricture from surgery. Once cholelithiasis is diagnosed cholecystectomy in children with SCD should be performed to avoid the morbidity associated such as cholecystitis, biliary colic, pancreatitis, bile duct obstruction and cholangitis. The SCD child with gallstones and bile duct obstruction, bile duct dilatation in US, pancreatitis, elevated total bilirubin and alkaline phosphatase or cholangitis should undergo an MRCP to rule out choledocholithiasis. If bile duct stones are diagnosed, then an ERCP with sphincterotomy and stone extraction is the next step in management. The advantage of the preoperative ERCP is that the bile ducts obstruction is cleared reducing chance of biliary infection. This is then followed by laparoscopic cholecystectomy. Should ERCP fails to clear the common bile duct of stones and the obstruction persists laparoscopic cholecystectomy with exploration of the bile duct and stone retrieval is in order. Recurrent biliary tract disease is a frequent complication of SCD (20% by age 4 years) and often presents as common bile duct obstruction by stone despite cholecystectomy. Increased perioperative complications may result from vaso-occlusion after transient hypoxia, hypothermia, dehydration and acidosis. Inadequate postoperative pain management may reduce respiratory effort leading to poor pulmonary toilet, hypoxia and acute chest syndrome from intrapulmonary vaso-occlusive crisis.

References:

- 1- Sandoval C, Stringel G, Ozkaynak MF, Tugal O, Jayabose S: Perioperative management in children with sickle cell disease undergoing laparoscopic surgery. *JLS*. 6(1):29-33, 2002
- 2- Vicari P, Gil MV, Cavalheiro Rde C, Figueiredo MS: Multiple primary choledocholithiasis in sickle cell disease. *Intern Med*. 47(24):2169-70, 2008
- 3- Ingle SS, Ubale P: Anesthetic management of a patient with sickle cell disease for common bile duct exploration. *J Anaesthesiol Clin Pharmacol*. 27(4): 547-549, 2011
- 4- Issa H, Al-Salem AH: Role of ERCP in the era of laparoscopic cholecystectomy for the evaluation of choledocholithiasis in sickle cell anemia. *World J Gastroenterol*. 17(14):1844-7, 2011

5- Al-Salem AH, Issa H: Laparoscopic cholecystectomy in children with sickle cell anemia and the role of ERCP. *Surg Laparosc Endosc Percutan Tech.* 22(2):139-42, 2012

6- Amoako MO, Casella JF, Strouse JJ: High rates of recurrent biliary tract obstruction in children with sickle cell disease. *Pediatr Blood Cancer.* 60(4):650-2, 2013

Congenital Uterovaginal Prolapse

Uterovaginal prolapse (UVP) occurring in the neonatal period is very rare. Uterovaginal prolapse refers to the downward descent with protrusion of the uterus and vagina to the exterior through the introitus. Uterovaginal prolapse occurs due to weakness of the cardinal ligaments and uterosacral ligaments that provide support to the uterus and vagina. Most cases are associated with congenital anomalies of the spine such as spina bifida occulta or myelomeningocele due to maldevelopment of sacral innervation to the levator ani muscle (perineal branch of the sacral nerve). Congenital UVP is also seen in babies with congenital cutis laxa. Congenital UVP manifest symptoms at birth or within the first few days of life. A few cases are caused by abnormal stress such as prolonged labor in breach condition, birth trauma or abnormality of the cervix. Diagnosis is clinical: pink-fleshy mass is seen protruding from the introitus with circumferential prolapse of the vaginal wall. The external cervical os is usually seen at the upper end of the lesion with a normal appearing urethral orifice. Differential diagnosis includes vaginal polyp, urethral prolapse, paraurethral cysts and rhabdomyosarcoma botryoides. CT or MRI is diagnostic and helps clear the differential diagnosis. Management of congenital uterovaginal prolapse is conservative with single or repeated manual reduction of the defect. Hypertonic saline packs or vaginal pessaries have also been utilized. As genital edema subsides the prolapse is usually maintained in place. Surgical intervention is indicated when conservative management fails, when the prolapse becomes recurrent after repeated reductions or when there is evidence of vaginal mucosal hypertrophy or ulceration. These include fixation of the uterus anteriorly to the urinary bladder with nonabsorbable sutures using a small suprapubic incision, sling or sacral cervicopexy. Inflated Foley catheter into the vaginal orifice after reduction for a period of two weeks can also successfully correct the uterovaginal prolapse.

References:

1- Abdelsalam SE, Desouki NM, Abd alaal NA: Use of Foley catheter for management of neonatal genital prolapse: case report and review of the literature. *J Pediatr Surg.* 41(2):449-52, 2006

2- Hyginus EO, John CO: Congenital uterovaginal prolapse present at birth. *J Surg Tech Case Rep.* 5(2):89-91, 2013

3- Jijo ZW, Betele MT, Ali AS: Congenital Uterovaginal Prolapse in a Newborn. *Case Rep Obstet Gynecol.* 2018 Jun 27;2018:1425953. doi: 10.1155/2018/1425953. eCollection 2018.

4- Saha DK, Hasan KM, Rahman SM, Majumder SK, Zahid MK, Chakraborty AK, Bari MS: Neonatal uterine prolapse - a case report. *Mymensingh Med J.* 23(2):401-5, 2014

5- Choudhary SV, Bisati S, Koley S: Congenital cutis laxa with rectal and uterovaginal prolapse. *Indian J Dermatol Venereol Leprol.* 77(3):321-4, 2011

6- Taksande AM, Vilhekar KY, Batra P, Jain M: Neonatal genital prolapse. *J Indian Med Assoc.* 109(7):502-3, 2011

Cushing's Syndrome

Cushing syndrome refers to the constellation of signs and symptoms associated with excessive production of cortisol. Cushing syndrome (CS) is caused by an ACTH producing pituitary or ectopic tumor (ACTH-dependent) or an adrenal adenoma/carcinoma (ACTH-independent). The child with CS develops hypertension, diabetes mellitus, hypokalemia, alkalosis, bone loss, fractures and psychiatric disturbances. CS patients suffer from infection, myocardial infarction or venous thrombosis. The primary source of ACTH should be eliminated as two-thirds of deaths are related to CS cortisol excess. Effective management includes normalization of cortisol levels or inhibiting steroid genesis to eliminate signs and symptoms of CS even if there is no established diagnosis. It also includes normalizing comorbidities using adjunctive therapy. Medical therapy with steroidogenesis inhibitors (metyrapone and ketoconazole) can reduce plasma cortisol levels. It is recommended that initial resection of primary lesions underlying Cushing disease, ectopic and adrenal etiology be performed to reduce glucocorticoid excess. These include resecting unilateral adrenal disease, ectopic ACTH-secreting tumors with node dissection, transsphenoidal selective removal of pituitary tumors and surgical resection of bilateral adrenal disorders. Most cases (90%) of pediatric CS are due to pituitary adenomas and they are primarily managed with trans-sphenoidal surgery. ACTH-independent causes of CS in children are less than 10% and comprises adrenal cortex adenomas or hyperplasia. Laparoscopic surgical resection of disease adrenal gland whether unilateral or bilateral is highly and rapidly effective in eliminating the source of cortisol. Infantile ACTH-independent CS is very rare and usually reported as part of McCune Albright (large adrenal glands due to macro-nodular hyperplasia) or Carney complex disease (pigmented nodular hyperplasia). Failed pituitary management of CS or selected patients with ACTH-dependent CS from unmanageable primary source with reasonable life expectancy may benefit from medical therapy with inhibitors plus bilateral removal of the adrenal glands for control of symptoms.

References:

- 1- Morris LF, Harris RS, Milton DR, et al: Impact and timing of bilateral adrenalectomy for refractory adrenocorticotrophic hormone-dependent Cushing's syndrome. *Surgery*. 154(6):1174-83, 2013
- 2- Nieman LK, Biller BM, Findling JW, et al: Treatment of Cushing's Syndrome: An Endocrine Society Clinical Practice Guideline. *J Clin Endocrinol Metab*. 100(8):2807-31, 2015
- 3- Paduraru DN, Nica A, Carsote M, Valea A: Adrenalectomy for Cushing's syndrome: do's and don'ts. *J Med Life*. 9(4):334-341, 2016
- 4- Simforoosh N, Razzaghy Azar M, Soltani MH, Nourbakhsh M, Shemshaki H: Staged Bilateral Laparoscopic Adrenalectomy for Infantile ACTH-independent Cushing's Syndrome (Bilateral Micronodular Non-pigmented Adrenal Hyperplasia): A Case Report. *Urol J*. 14(5):5030-5033, 2017
- 5- Maccora D, Walls GV, Sadler GP, Mihai R: Bilateral adrenalectomy: a review of 10 years' experience. *Ann R Coll Surg Engl*. 99(2):119-122, 2017
- 6- Al-Otaibi KM, Abduljabbar MA: Symptomatic cycling Cushing disease managed by simultaneous bilateral laparoscopic adrenalectomy in a 11-year-old boy. *J Family Community Med*. 21(1):61-4, 2014

***Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP**
 Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,

**Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Children's & Woman
Hospital.**

**Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-
0426.**

Tel (787) 340-1868 E-mail: *titolugo@coqui.net*

Internet: <http://home.coqui.net/titolugo>

**8 PSU 1993-2019
ISSN 1089-7739**