International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

Chronic Calcific Pancreatitis in A Child- Clinical Presentation, Diagnosis and Management.

Aastha Agnihotri[1], Komal Garg[2].

[1]Department of General Surgery, Indira Gandhi Medical College, Shimla, India.
[2]Department of Otorhinolaryngology, Indira Gandhi Medical College, Shimla, India.

ABSTRACT

Context Chronic Pancreatitis is a rare, but important disease in children and adolescents with negative impact on quality of life. It is necessary to recognise this condition as one of the differential diagnosis of recurrent epigastric pain and bringing together a multidisciplinary team for effective diagnosis and management.

Case Report A 16 year old male child with history of recurrent pain abdomen was diagnosed with chronic calcific pancreatitis with severe exocrine insufficiency with pain with duct centeric disease cause idiopathic, without any family history for which Puestow's procedure was performed.

Conclusion Successful treatment of chronic pancreatitis reduces the risk of life threatening exacerbations, allows the patient to have significant improvements in life quality, alleviating daily symptoms that may have been present for years.

KEYWORDS: Chronic pancreatitis, Puestow procedure, pancreaticojejunostomy, paediatric surgery

INTRODUCTION

Pancreatic disorders in paediatric population have been increasingly recognised in recent years but literature on chronic pancreatitis in children remains limited. It is characterised by progressive irreversible inflammatory destruction of the gland, leading to pancreatic fibrosis and ductal changes with subsequent loss of pancreatic exocrine and endocrine functions(1,2). The incidence of CP in children has been estimated at 2 cases per 100000 persons and prevalence has been reported as 5.8 per 1000,00 persons(3). Although the incidence is low but the overall disease burden is considerably high, with numerous emergency department visits, hospitalisation, procedures and interventions (4). There are multiple risk factors for CP in children with genetic causes accounting for majority of cases followed by obstructive and anatomical causes (5). Other identified risk factors include autoimmune and metabolic risk factors, hypertriglyceridemia, smoking, medications, alcohol etc (5). In the INSPPIRE (INternational Study Group of Pediatric Pancreatitis: In search for a cuRE) study, approximately 80% of children with CP complained of abdominal pain; pain was constant and requiring opioids in approximately 33% (5). Endocrine and exocrine pancreatic dysfunction are potential sequelae of disease progression.

Varying imaging modalities may be utilized to detect parenchymal changes and ductal abnormalities in the diagnosis of CP (6). Transabdominal ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), magnetic resonance cholangiopancreatopography(MRCP), and endoscopic ultrasound (EUS) are imaging studies incorporated in the CP work-up with varied sensitivities and specificities (7). Endoscopic retrograde cholangiopancreatography (ERCP) can detect the pancreatic ductal changes of CP

and provide a therapeutic approach via dilation and stenting of ductal strictures and removal of obstructing stones. ERCP is being used with increased frequency in the paediatric population with reported complication rates similar to adults (0%–11%) (1,8,9).

The primary goals of management of CP are control of pancreas-associated pain, treatment of exocrine and endocrine pancreatic insufficiency, and treatment of CP-related complications. Adult studies have reported that approximately 50% of CP patients will eventually require an operation for management of pain, complications, or suspicion of associated malignancy (10,12). Although similar data are not available in the paediatric population, surgery does play a role in the management of children with CP who do not have other medical or endoscopic options available. The most common indication for operation in CP is debilitating pain that has failed medical and endoscopic management, with less common indications in children including duodenal or biliary obstruction, pseudocysts, or suspicion of malignancy (13,15). The various surgical options may be divided into 2 categories: conventional operations (ie, drainage procedures, partial resections, and procedures combining drainage and resection); and total pancreactectomy with islet autotransplantation (TPIAT) (13 – 16). Selection of an operation must take into account anatomic and morphologic features of the pancreas, such as main duct dilation and/or the presence of an inflammatory head mass. The presence of underlying genetic risk factors for pancreatitis may also impact operation. Because of the paucity of literature on operations for CP in children, paediatric management has largely been based on adult data. The goals of this document are to summarise the existing literature regarding the surgical management of CP in children, to provide recommendations for surgical management in children with CP based on available paediatric and adult data, and to identify knowledge gaps for future studies. Medical and endoscopic management o CP based on available paediatric and adult data, and to identify knowledge gaps for future studies. Medical and endoscopic management of CP have been covered in 2 separate documents prepared by the Pancreas committee of NASPGHAN (17,18).f CP have been covered in 2 separate documents prepared by the Pancreas committee of NASPGHAN (17,18).

CASE REPORT

A 16 year old male child, resident of Shimla, Himachal Pradesh, India, presented with complaints of pain in upper abdomen 4 yrs back which was sudden in onset, radiating to back, severe, boring in character, relieved on bending forward and increased on movement. Patient took treatment from local hospital and was relieved. Similar episode occurred 2 weeks back which was also associated with vomiting, 5-6 episodes per day, non projectile, bilious, watery content which was relieved after symptomatic treatment was given in paediatrics department, later after radiological investigations, he was referred to surgery department.History of loss of appetite was present. No history of fever, jaundice, loose motions, lump abdomen.

On examination the patient was of thin built. General physical examination was normal. Per abdomen examination tenderness was present in epigastric region, rest was normal. Routine haematological investigations showed raised serum lipase(118.4), raised serum ALP(243), rest of investigations were normal.

USG abdomen showed multiple cysts along main pancreatic duct with diameter of cysts upto 16mm.pancreatic tissue appears to be reduced and appears to be grossly replaced with these cysts. There are multiple echogenic foci in these cysts/pancreatic duct, the residual pancreatic tissue shows increased echogenicity. **MRCP** showed Gall bladder with Phrygian cap, pancreatic duct measures 17mm, dilated

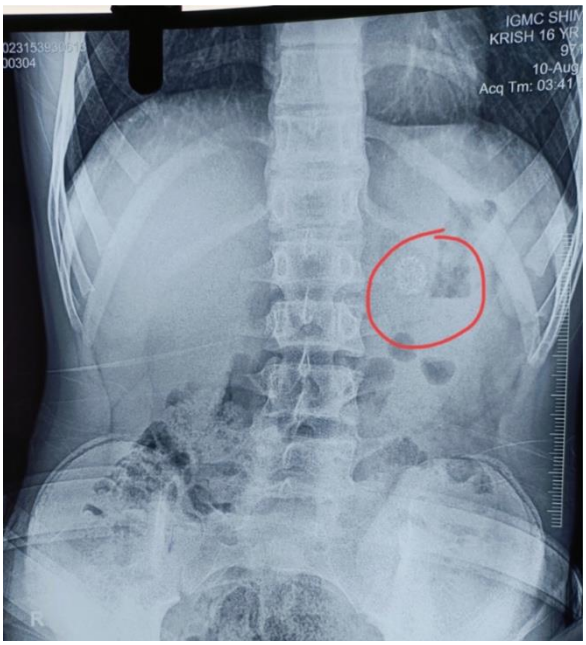
and tortuous. There is presence of multiple large signal voids seen in pancreatic duct in head and tail region largest 2.8x2cm in tail region. Multiple small outpouching are also seen adjoining the ducts. MPD is seen inserting into ampulla separately and is seen only for segment of 1.4cm from ampulla. Pancreas is not visualised likely atrophied.

Hrct thorax and CFTR gene analysis was normal. Fecal elastase was 14.8ug/g stool

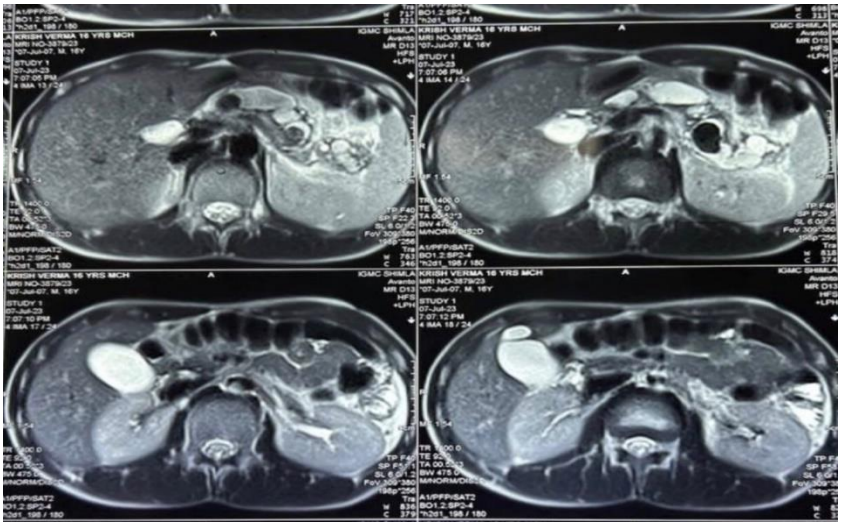
Diagnosis of "chronic calcific pancreatitis with severe exocrine insufficiency with pain with duct centeric disease cause idiopathic, without any family history" was made.

Patient was planned for Modified Puestow's procedure with Roux-En-Y Pancreaticojejunostomy with mesentric lymph node and pancreatic tissue biopsy.

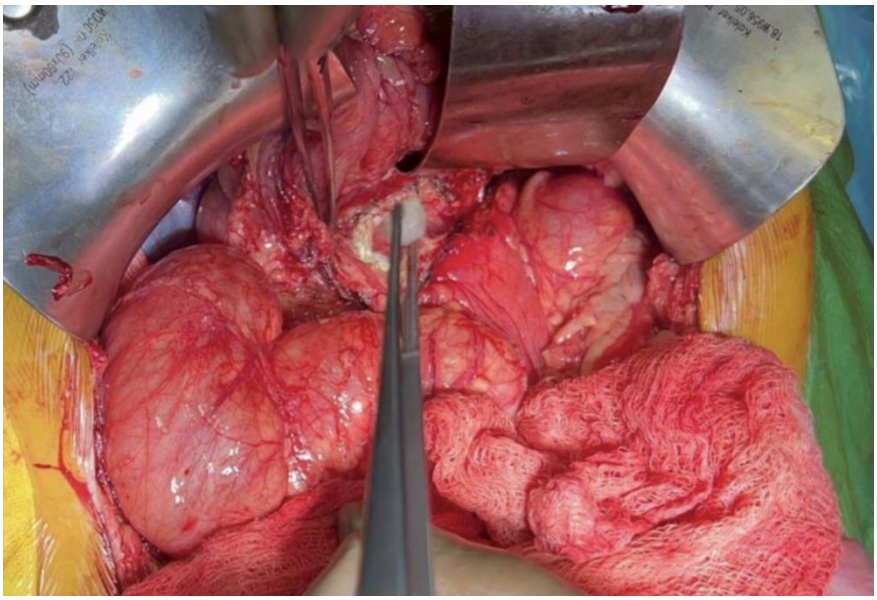
Intaoperatively MPD was 2cm. 2 large calculi in neck and tail with multiple concretions were seen, spiral like valve was present in MPD, body and neck. Pancreas in body , tail and neck was hard and firm. On Post operative day 3, drain fluid amylase was raised- grade 1 post operative pancreatic fistula which later came within normal limit. Patient was discharged on post operative day 8 with follow up.



Abdominal X-ray showing radio-opaque shadow in pancreatic region



MRCP showing multiple large signal voids seen in pancreatic duct in head and tail region



Intra operative picture showing retrieval of stone from dilated main pancreatic duct.

DISCUSSION

The most common indication for surgical intervention in CP is the persistent pain impacting quality of life that is not adequately relieved by medications or endoscopic approaches. The ultimate goal of operation is to provide pain relief and liberate the patient from opioids, while preserving endocrine and exocrine pancreatic function. There are only a few reports of surgical interventions in children with CP, derived from small cohorts of patients without long-term out- comes. Therefore, it is not known how frequently surgical interventions are utilised or whether certain surgical procedures are indicated over others in children with CP. Overall, it is estimated that up to 50% of patients with CP will require operation during their lifetime (11,12,14,19). In the INSPPIRE cohort, at least 1 pancreatic operation was performed in 39% of children, including longitudinal pancreaticojejunostomy in 14%, partial resection in 1%, and TPIAT in 28% (4). It must be recognised, however, that INSPPIRE is a research consortium, and these findings may not be representative of the overall distribution of clinical cases of CP in children.

Caring for children with CP requires a multidisciplinary approach with a team that consists of pancreatologists, endoscopists, surgeons, radiologists, pain specialists, psychologists, endocrinologists, and nutritionists. The specific operation must be tailored to the anatomy and morphology of pancreatic disease, and not 1 single surgical option can be recommended for every CP patient. Conventional surgical approaches are classified into the following groups: drainage operations to decompress a dilated ductal system, resections to remove a portion of the pancreas (most commonly, an inflammatory mass in the head of the pancreas), or a combination of drainage and resection (eg, duodenum-preserving pancreatic head resections, including Beger procedure, Berne procedure, and Frey procedure). Total pancreatectomy with islet autotransplantation is a surgical option in patients who do not have the anatomy or morphology of disease to consider a drainage operation or partial resection and in patients who have failed previous conventional surgical approaches, or when genetic predispositions are present. In the latter cohort, the total pancreatectomy definitively addresses the underlying ‘‘field defect’’ of a genetic risk factor. We have developed an algorithm to determine the optimal surgical approach for children with CP that has failed all medical or endoscopic approaches.

During a Puestow procedure, also known as a lateral pancreaticojejunostomy, the abdomen is opened with an incision from the lower breastbone to the belly button. The pancreas is exposed and the main pancreatic duct is opened from the head to the tail of the pancreas. The opened pancreatic duct is then connected to a loop of small intestine so that the pancreas drains directly into the intestines. This procedure has been done with much success in treating pain associated with chronic pancreatitis for over 50 years. With a success rate of 70 - 90 percent, this procedure has a low operative complication rate and a very low mortality rate.

CONCLUSION

Disease morphology and genetic risk factors must be considered while determining the optimal surgical approach for children with chronic pancreatitis. Outcome can differ from case to case and type of intervention being done. A multidisciplinary team approach is needed to assure the best possible management for each patient.

Conflict of interest No competing or financial interests

References

1. Taylor CJ, Chen K, Horvath K, et al. ESPGHAN and NASPGHAN Report on the assessment of exocrine pancreatic function and pancrea- titis in children. J Pediatr Gastroenterol Nutr 2015;61:144 – 53.
2. Spanier B, Bruno MJ, Dijkgraaf MG. Incidence and mortality of acute and chronic pancreatitis in the Netherlands: a nationwide record-linked cohort study for the years 1995 – 2005. World J Gastroenterol 2013;19:3018 – 26.
3. Sellers ZM, MacIsaac D, Yu H, et al. Nationwide trends in acute and chronic pancreatitis among privately insured children and non-elderly adults in the United States, 2007 – 2014. Gastroenterology 2018;155:469.e1 – 78.e1.
4. Schwarzenberg SJ, Bellin M, Husain SZ, et al. Pediatric chronic pancreatitis is associated with genetic risk factors and substantial disease burden. J Pediatr 2015;166:890.e1 – 6.e1.
5. Kumar S, Ooi CY, Werlin S, et al. Risk factors associated with pediatric acute recurrent and chronic pancreatitis: lessons from INSPPIRE. JAMA Pediatr 2016;170:562–9.
6. Della Corte C, Faraci S, Majo F, et al. Pancreatic disorders in children:
7. new clues on the horizon. Dig Liver Dis 2018;50:886 – 93.
8. Mayerle J, Hoffmeister A, Werner J, et al. Chronic pancreatitis– definition, etiology, investigation and treatment. Dtsch Arztebl Int 2013;110:387 – 93.
9. Issa H, Al-Haddad A, Al-Salem AH. Diagnostic and therapeutic ERCP
10. in the pediatric age group. Pediatr Surg Int 2007;23:111 – 6.
11. Darge K, Anupindi S. Pancreatitis and the role of US, MRCP and ERCP. Pediatr Radiol 2009;39(Suppl 2)S153-7.
12. Steer ML, Waxman I, Freedman S. Chronic pancreatitis. N Engl J Med 1995;332:1482 – 90.
13. Buchler MW, Friess H, Muller MW, et al. Randomized trial of duode- num-preserving pancreatic head resection versus pylorus-preserving Whipple in chronic pancreatitis. Am J Surg 1995;169:65 – 9.
14. Mitchell RM, Byrne MF, Baillie J. Pancreatitis. Lancet 2003;361:1447– 55.
15. Parekh D, Natarajan S. Surgical management of chronic pancreatitis. Indian J Surg 2015;77:453 – 69.

16. Issa Y, van Santvoort HC, van Goor H, et al. Surgical and endoscopic treatment of pain in chronic pancreatitis: a multidisciplinary update. Dig
17. Surg 2013;30:35 – 50.
18. van der Gaag NA, Gouma DJ, van Gulik TM, et al. Review article: surgical management of chronic pancreatitis. Aliment Pharmacol Ther 2007;26(Suppl 2):221 – 32.
19. Strobel O, Buchler MW, Werner J. Surgical therapy of chronic pan- creatitis: indications, techniques and results. Int J Surg 2009;7:305 – 12.
20. Surgery in Children With Chronic Pancreatitis
21. Liu QY, Gugig R, Troendle DM, et al. The roles of endoscopic ultrasound and endoscopic retrograde cholangiopancreatography in the evaluation and treatment of chronic pancreatitis in children: a position paper from the North American Society for Pediatric Gastro- enterology, Hepatology, and Nutrition Pancreas Committee. J Pediatr Gastroenterol Nutr 2020;70:681–93.
22. Freeman AJ, Maqbool A, Bellin MD, et al. Medical management of chronic pancreatitis in children: a position paper by the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition Pancreas Committee. J Pediatr Gastroenterol Nutr 2021;72:324 – 40.
23. Ray S, Sanyal S, Ghatak S, et al. Frey procedure for chronic pan- creatitis in children: a single center experience. J Pediatr Surg 2015;50:1850 – 3.