

Extracorporeal shock wave lithotripsy in the management of a 14-year-old girl with chronic calcific pancreatitis

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ABSTRACT

Background. Chronic pancreatitis is very rare in childhood and causes chronic/relapsing abdominal pain, frequent hospitalizations, malnutrition, growth retardation, and stone formation in the main duct. Although pancreatic extracorporeal shock wave lithotripsy (P-ESWL) is commonly used in the treatment of pancreatic stones (PS) in adults, the use in children is still controversial. An adolescent girl with multiple PS is presented to discuss the use of ESWL as a treatment alternative in children with PS.

Case. A 14-year-old girl was admitted with abdominal pain and elevated pancreatic enzyme levels. Abdominal US showed irregularity and rough echogenicity in pancreas revealing pancreatitis. Multiple stones were seen in main pancreatic duct on Magnetic resonance cholangiopancreatography (MRCP). Endoscopic retrograde cholangiopancreatography (ERCP) was performed and dilated pancreatic duct, thickened pancreatic secretion were detected. Endoscopic sphincterotomy was performed. Endoscopic removal of stones could not be achieved since the largest stone was 17x8 mm. Pancreatic extracorporeal shock wave lithotripsy (P-ESWL) was performed using electromagnetic lithotripter under general anesthesia. Following ESWL, fragmentation of stones in the main duct was confirmed with ERCP. After 3 sessions of ESWL, no ESWL-related complication was observed. Pain relief was achieved. The patient is still under follow-up regarding endocrine and exocrine function of pancreas.

Conclusion. ESWL may be an effective and safe management option in pediatric PS which could not be removed by ERCP. The patients managed with ESWL should be followed-up for a long time regarding the endocrine and exocrine functions of the pancreas. As in management of adult pancreatitis, clinical guidelines are needed regarding the management of pediatric PS.

Key words: calcific pancreatitis, chronic pancreatitis, ESWL, pancreatic stone, children.

Chronic pancreatitis (CP) is rarely seen in children and causes extreme disruption not only in their health status but also in their education and development.¹ CP causes chronic and relapsing abdominal pain, frequent hospitalizations, malnutrition and growth retardation in children.¹ Approximately 50% of

CP patients suffer from pancreatic ductal stones which increase intra-ductal pressure and cause severe abdominal pain.^{1,2} In addition, children have increased cumulative risk of recurrence since their life expectancy is longer than adults. Therefore, treatment of choice should be minimal invasive as possible to ensure repetitive procedures could be done safely in children.

Although pancreatic stone (PS) management is considered as standard treatment in adults with pancreatic extracorporeal shock wave lithotripsy (P-ESWL), there is no consensus on management of PS in children. ESWL was first used for renal calculi by Chaussy et al.³ in

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1980, and its use for pancreatic stones was by Sauerbrunch et al.⁴ in 1987. Since then, it has become the first line therapy in PS management in adults as recommended by the European Society of Gastrointestinal Endoscopy (ESGE) guideline.⁵ However, in the same guideline it was reported that endoscopic management should be the treatment of choice in children since effectiveness and safety of ESWL use in children was not yet verified.⁵ Although a few studies have reported the use of P-ESWL in children in ESGE guideline, there is no standardized protocol or a guideline regarding use of P-ESWL in children. Herein, we reported a child with multiple PS who was treated with P-ESWL to discuss the management of PS in children.

Case Report

A 14-year-old girl was admitted to our department with abdominal pain starting from the epigastric region and surrounding her belly. Her physical examination revealed epigastric tenderness. The weight of the patient was 42 kg (10-25 percentiles) and height was 150 cm (5 percentiles). The laboratory work-up were normal except amylase and pancreatic amylase levels. The amylase level was 1621 U/L (N: 28-100 U/L) and pancreatic amylase level was 1216 U/L (N: 13-53 U/L). Abdominal ultrasonography (US) showed irregularity and rough echogenicity in the pancreas revealing

pancreatitis. Therefore, she was hospitalized and medical treatment was initiated.

In her past medical history, she experienced three attacks of pancreatitis starting from 9 years of age. During the second episode of pancreatitis, endoscopic retrograde cholangiopancreatography (ERCP) was performed and partial divisum, dilated pancreatic duct, and thickened pancreatic secretions were detected. Endoscopic sphincterotomy was performed, but her complaints recurred. The blood lipid levels, serum chemistry, liver function tests, thyroid function tests and genetic analysis for cystic fibrosis were within normal limits.

At her present admission, multiple stones were seen in the main pancreatic duct on Magnetic resonance cholangiopancreatography (MRCP) (Fig. 1). Endoscopic removal of stones could not be achieved since the largest stone was 17x8 mm. Endoscopic sphincterotomy was performed again and our multidisciplinary council decided to perform a P-ESWL. P-ESWL was performed using a third-generation electromagnetic lithotripter (Siemens modularis variostar-Cplus, Erlangen, Germany) under general anesthesia. PS were targeted by lithotripter using fluoroscopy (Fig. 2). Not more than 5000 shocks with 3.5 microjoule per each shock were performed for each ESWL session (16 000 kV, 120 shock/min). The duration of each session was approximately 60 min. P-ESWL sessions were

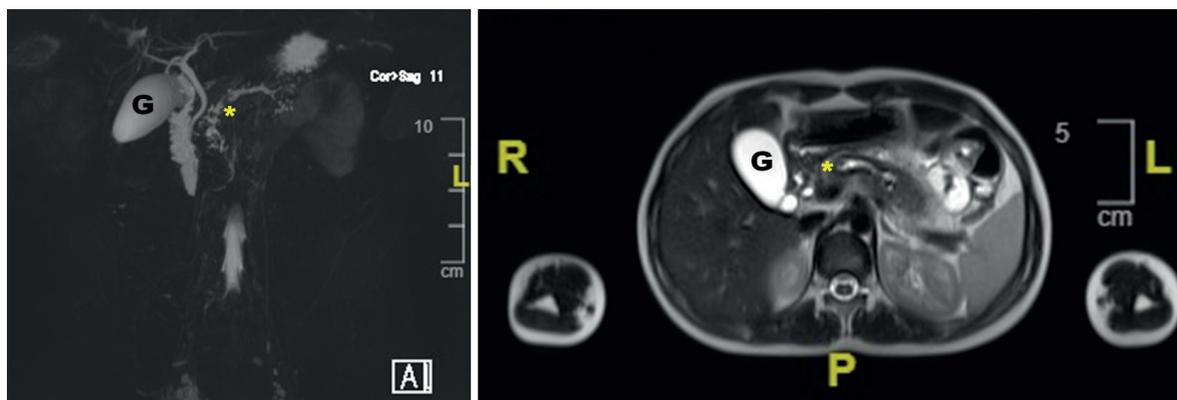


Fig. 1. The MRCP images showing dilated and tortoise main pancreatic duct with irregular borders and multiple stones within the main pancreatic duct (G: gallbladder, *: main pancreatic duct).

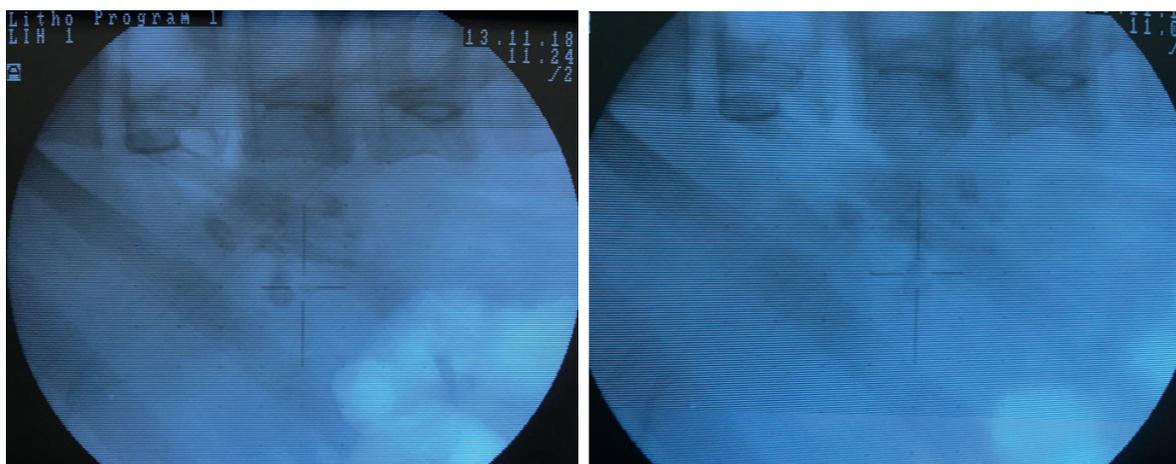


Fig. 2. Pancreatic stones were targeted by lithotripter using fluoroscopy showing a large stone at the middle of the target lines.

performed every other day, three times totally. Radiation protection was provided according to ESGE guidelines.⁵ Following ESWL, ERCP was performed showing that the stones were fragmented and passage of contrast agent could be achieved into the main pancreatic duct. Since spontaneous clearance of stones was not seen, endoscopic wash of the pancreatic duct and re-sphincterotomy was performed. Additionally, 7Fr stent was inserted into the main pancreatic canal to prevent re-obstruction of the canal. After P-ESWL sessions, no ESWL-related complication was observed. Pain relief could be achieved with non-steroidal anti-inflammatory drugs in small doses.

Abdominal US was performed 1 month after the last ESWL session and revealed fragmented stones in pancreatic duct with a largest diameter of 7 mm. Therefore, we decided to repeat the P-ESWL sessions 3 months later. The pancreatic exocrine function profile was evaluated with testing for steatorrhea and fecal elastase test. Although steatorrhea in the stool was negative, fecal elastase level was lower than 15 mg/ml revealing pancreatic exocrine function deficiency. Therefore, pancreatic enzyme replacement 4 times/day (Creon®, Abbott Ltd., Istanbul, Turkey; 25000- 300 mg: 300 mg pancreatin + 18000 PEU amylase + 25000 PEU lipase + 1000 PEU protease) was ordered. Her vitamin D (Vit D) level was 5,4 µg/l (severe

deficiency) and Vit D replacement (50000 IU/15 ml, 6 drop/day) was ordered. The Vit D level increased to 20,19 µg/l after replacement therapy. The pancreatic endocrine function was assessed with blood glucose level, 75-g oral glucose tolerance test and HbA1c level which were within normal limits. The patient is still under follow-up for 1 year regarding endocrine and exocrine function of pancreas. The current weight of the patient is 46 kg (10-25 percentiles) and height is 155 cm (10-25 percentiles) revealing a slow improvement. In addition, she still has a stent in the main pancreatic canal with fragmented and partially cleared stones. Since then, she has not experienced pancreatitis again. Informed consent was obtained from the parents of the patient that allows using the clinical data for scientific purposes.

Discussion

Pancreatic duct stones (PS) are seen in 50% of patients with chronic pancreatitis. The intraductal pressure increases in the presence of PS leading to ischemia and pain.^{1,2} Since life expectancy of children is longer, life-long risk for PS recurrence is higher in children. The relapsing abdominal pain causes frequent admissions to emergency departments and longer hospital stays leading to increased school absences. Therefore, repeatable, safe

and effective methods are needed in the management of PS in children.

Although PS management is standardized in adults, there is no consensus on management of PS in children. The treatment alternatives of PS in children include surgical excision, endoscopic removal or fragmentation of stones by ESWL. Endoscopic stone removal is the first line treatment since it is less invasive than surgery. However, success rate of endoscopy is limited to 50% of all PS in children.^{1,2} Additionally, the PS smaller than 3 mm and located at the head and body of the pancreas are considered as an indication for ERCP. The stones greater than 5 mm, multiple in number and distal location cannot be removed by ERCP. Therefore, fragmentation of such stones is needed to restore pancreatic juice flow and relieve pain.²

P-ESWL is reported to be safe, effective and minimal invasive in adults and recommended as first-line therapy combined with or without endoscopic removal of fragments.⁵ The success in P-ESWL is defined as fragmentation of stones into pieces smaller than 3 mm or decreased stone density in radiological examinations.⁵ A meta-analysis revealed that complete ductal clearance is achieved in 70% of cases and partial ductal clearance is achieved in 22% of cases.⁶ According to the results in that meta-analysis, number of sessions varied according to the clinical response and ductal clearance, but better results were obtained when the number of sessions were more than three.^{2,6} Additional endoscopic procedures such as removal of fragments, stent placement to main pancreatic duct and sphincterotomy are usually needed following ESWL.^{5,6} In our case, partial ductal clearance, fragmentation of stones and pain relief were achieved after 3 sessions of ESWL. Endoscopic sphincterotomy was performed to increase flow of pancreatic juice; however, it was not possible to place a stent because of multiple fragments in the entire duct. Additional sessions were planned since the patient has residual stones in pancreatic duct.

The endocrine and exocrine function of the pancreas should be investigated in patients with chronic pancreatitis.⁷ Although there is not enough data in the literature, ESWL seems to improve exocrine and endocrine pancreatic functions.^{6,7} When we evaluated the endocrine and exocrine pancreatic function in this patient, we found decreased exocrine function with normal endocrine functions. However, this assessment was performed only after ESWL; we cannot comment on the baseline evaluations since we were the final referral center for this patient. Therefore, it will not be possible to suggest that ESWL either improves or alters the exocrine or endocrine pancreatic function for the present case. Thus, all clinicians should be aware of alterations in pancreatic functions in cases with chronic pancreatitis and follow-up the result of the management. On the contrary, Adamek et al.⁸ stated that endoscopic management or ESWL do not affect the development of glandular insufficiency, since chronic pancreatitis causes progressive parenchymal destruction of pancreas. Therefore, future studies are needed to investigate the effect of management of chronic pancreatitis and PS to pancreatic functions in children.

Although P-ESWL was reported to be safe and effective in the management of PS, it may cause several complications including acute pancreatitis, bleeding, infection or perforation.¹ Wang et al.¹ reported that complication rates were 11% in children and 12% in adults and they found no statistically significant difference between pediatric and adult patients. The present case did not experience any complications after the P-ESWL sessions. Although future studies are needed to confirm these results, P-ESWL in children seems to have similar complication rates with adult patients and can be performed safely in childhood PS management.

In conclusion, ESWL is an effective and safe management option for pediatric PS that could not be removed by ERCP. The patients managed with ESWL should be followed-up for

a long time in regards to endocrine and exocrine functions of the pancreas. As in management of adult pancreatitis, clinical guidelines are needed regarding the management of pediatric PS.

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