HYPERTRIGLYCERIDAEMIA INDUCED ACUTE PANCREATITIS IN AN INFANT: A CASE REPORT

Dr. Amna Barghout* and Abdulmoein Eid Al-Agha, MBBS, DCH, CABP, FRCPCH
King Abdul-aziz University Hospital.

*Corresponding Author: Dr. Amna Barghout
King Abdul-aziz University Hospital.

ABSTRACT
Hypertriglyceridaemia is a rare cause of acute pancreatitis in children, which requires a high index of clinical suspicion. We report a case of a 6-month-old male infant who presented to the emergency department with the acute onset of vomiting, nausea, irritability, and inconsolable crying. He was moderately dehydrated, with generalized abdominal distension and moderate tenderness. His laboratory workup revealed hypertriglyceridaemia, normal cholesterol levels, and elevated lipase and amylase levels. Based on this case, we suggest that abdominal US should be performed in any case of suspected pancreatitis or unexplained abdominal pain.

KEYWORDS: Acute, pancreatitis, hypertriglyceridaemia, amylase, infant.

INTRODUCTION
The incidence of pancreatitis in children has increased significantly in the past two decades. It is estimated that 2 to 13 new cases occur annually per 100,000 children. Although the pathophysiology and functional consequences in children are identical to adults, its aetiology differs significantly. The most common causes of pancreatitis in children include systemic diseases, such as systemic lupus erythematosus, Henoch-Schönlein purpura, Kawasaki disease, and Crohn’s disease, hypolipoproteinaemia, hypertriglyceridaemia (HTG), various drugs, toxins, infections, trauma, and hereditary pancreatitis.

The association between hyperlipidaemia and acute pancreatitis was first noted by Speck in 1865. Since then, the correlation has been studied by many researchers, most importantly by Cameron and associates. HTG typically presents with acute or recurrent subacute pancreatitis. The chronic form is rare. Clinical indicators of acute pancreatitis include the sudden onset of sharp, persistent abdominal pain associated with nausea, vomiting, and fever. Additionally, the presence of hyperamylasaemia accompanied by a compatible clinical presentation is needed to confirm the diagnosis of acute pancreatitis.

Diagnostic accuracy is further improved if there is an elevated serum lipase level (99% specificity). Serum lipase levels may remain elevated, while amylase levels return to normal. We report a case of primary familial hypertriglyceridaemia with secondary acute pancreatitis in a male infant.

CASE REPORT
Consent to report this case was obtained from the infant’s father.

A 6-month-old male infant presented to the emergency department with the acute onset of vomiting, nausea, irritability, and inconsolable crying. On examination, he was conscious, alert, and irritable with moderate signs of dehydration. His growth parameters were as follows: length, 67 cm (50th percentile); weight, 6 kg (50th percentile). Respiratory rate was 30 breaths/min, pulse rate was 140 beats/min, blood pressure was 90/62 mmHg, and temperature was 37°C (axillary). The abdomen was moderately distended with generalised moderate tenderness. His laboratory workup revealed hypertriglyceridaemia, normal cholesterol levels, and elevated lipase and amylase levels. Based on this case, we suggest that abdominal US should be performed in any case of suspected pancreatitis or unexplained abdominal pain.

Abdominal ultrasonography (US) showed an enlarged echogenic heterogeneous texture of the pancreas with no dilation of the pancreatic duct, confirming acute pancreatitis with no obvious pseudocyst formation (Figure 1).
Table 1: Laboratory values showing hypertriglyceridaemia and elevated pancreatic enzymes.

<table>
<thead>
<tr>
<th>Serum tests</th>
<th>On Admission</th>
<th>On Discharge</th>
<th>Reference value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Triglyceride</td>
<td>27.33</td>
<td>6.61</td>
<td>0.30-2.30 mmol/L</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>3.34</td>
<td>1.48</td>
<td>0.5.20 mmol/L</td>
</tr>
<tr>
<td>Low density lipoprotein</td>
<td>1.26</td>
<td>0.680</td>
<td>0.3.57 mmol/L</td>
</tr>
<tr>
<td>Amylase</td>
<td>50</td>
<td>10</td>
<td>25-115 mg/dl</td>
</tr>
<tr>
<td>Lipase</td>
<td>1116</td>
<td>381</td>
<td>73-393 mg/dl</td>
</tr>
</tbody>
</table>

Figure 1: Ultrasound showing mild thickening of the pancreas.

During hospitalization, he was kept nil per mouth and given fluid hydration alongside analgesia, omega 3 fatty acids, niacin, and fibrate therapy. The patient resumed oral feeding within 2 days after cessation of vomiting and improved tenderness. He was discharged in good health.

DISCUSSION
Pancreatitis has been identified as the most common pancreatic disorder in children in recent reviews by Synn et al.\(^8\) and Vane et al.\(^9\) Hypertriglyceridaemia is a rare cause, contributing to acute pancreatitis in up to 7% of cases.\(^10\) The patient presented here developed acute pancreatitis secondary to hypertriglyceridaemia, a rare cause that can frequently lead to misdiagnoses. In the presence of a family history of familial hypertriglyceridaemia, high triglyceride levels will be present on laboratory analysis. However, the diagnosis of pancreatitis can usually be made with reasonable certainty based on clinical, radiographic, and laboratory findings.\(^11\) In this case, the patients presented with vomiting, nausea, irritability, and abdominal tenderness. Further diagnostic evaluations included an abdominal US. Abdominal US has been shown to have 80% accuracy in the evaluation of pancreatitis, usually demonstrating decreased echogenicity of the pancreas.\(^12\) The laboratory findings at admission showed elevated triglycerides, amylase, and lipase. In most reported cases, levels of liver enzymes, bilirubin, and amylase are either normal or slightly elevated.\(^12\) While pancreatitis rarely occurs except when triglyceride levels exceed 1500 mg/dL,\(^13\) mild to moderately elevated levels of triglycerides (200-1000 mg/dL) can usually be detected in the early stages.\(^14\) Janowitz considered serum amylase to be the most important diagnostic aid in determining pancreatic injury.\(^15\) Although serum amylase level may be normal in pancreatitis,\(^16\) up to 95% of cases of acute pancreatitis have elevated amylase levels.\(^17\) The standard management of pancreatitis consists of bowel rest with fluid hydration, with or without nasogastric suction. In 30% to 76% of cases, paediatric pancreatitis can be treated conservatively.\(^18\) Isolated cases of pancreatitis due to hypertriglyceridaemia have been reported and treated with plasma and lipoprotein apheresis with favourable outcomes; this therapy was not applicable to this case as a body weight greater than 20 kg is needed to perform apheresis. Octreotide, a somatostatin analogue, has also been used because it activates receptors in the pancreas that modulate pancreatic exocrine secretion.\(^19\) The mortality in paediatric pancreatitis varies greatly (0% - 78%).\(^20\) Nearly one-quarter of children with acute pancreatitis develop a severe complication, and the mortality rate in these instances is approximately 4%, despite significant advances in the treatment of this disease.\(^21\)

In our patient, the presence of a family history of hypertriglyceridaemia, with normal laboratory values of cholesterol and low density lipoprotein (LDL), led us to the diagnosis of type IV hypolipoproteinaemia. Screening result of the family was negative with positive consanguinity.
Familial hypertriglyceridaemia (FHTG) is an autosomal dominant disorder occurring in approximately 1 per 500 persons. In FHTG, the serum triglyceride levels range from 250-1000 mg/dl, with normal to mildly increased cholesterol levels. The molecular aetiology of this disorder of lipoprotein metabolism is unknown and the metabolic basis is heterogeneous.\(^{[22]}\)

FHTG, a type IV hyperlipidaemia, is exacerbated by sedentary lifestyle, obesity, insulin resistance, and oestrogen use. The heterogeneous metabolic basis includes impaired catabolism of triglyceride-rich lipoproteins, over-production of VLDL by the liver (hence overloading the normal catabolic processes), and a genetic overproduction of apo C-III.\(^{[23]}\) The apo A-I and apo A-II levels and the ratio of apo A-I to apo A-II are decreased.\(^{[23]}\) The diagnosis is confirmed when triglycerides are elevated (>90th percentile) with normal or mildly increased cholesterol (<90th percentile). FHTG disease is usually present in at least one first degree relative; in the present case, only the patient was affected. Complications are rare, with some patients presenting with pancreatitis and serum triglyceride level above 2000 mg/dl, as in the current case. The risk of coronary artery disease is quite low.\(^{[22]}\)

The mainstay of treatment for hypertriglyceridaemic pancreatitis consists of fibrates, which reduce plasma triglyceride levels by up to 50% and raise high-density. Fibrates modulate peroxisome %20einos (HDL) cholesterol levels by lipoproprotulator activated receptors-α (PPAR-α) in the liver, which decreases hepatic. They also 2411secretion of VLDL and increased lipolysis of plasma triglycerides [fatty acids -3]. Omega-3 increase HDLparticles reduce small dense LDL(eicosapentanoic and docosahexanoic acid) reduce plasma triglycerides by 20% when \(\mbox{fatty acids -3, } \mbox{Omega-3 increase HDL particles reduce small dense LDL(eicosapentanoic and docosahexanoic acid) reduce plasma triglycerides by 20% when } \]. Antioxidant 54lowering therapies [2-used in combination with other triglyceridetherapies (selenium, \(\mbox{β} \) carotene, vitamin C, \(\mbox{α} \)-tocopherol) have been used in the reduction of recurrent pancreatitis episodes in patients that remain markedly hypertriglyceridaemic after medical therapy, by virtue of their protection from free radical-induced acinar damage.

CONCLUSION
We report a new case of acute pancreatitis secondary to hypertriglyceridaemia. Acute pancreatitis should be highly suspected in any children who present with vomiting and nausea with abdominal pain and high lipid content. Our case supports the idea that abdominal US should be performed in any case of suspected pancreatitis or unexplained abdominal pain.

REFERENCES
10. Gan SI, Edwards AL, Symonds CJ. Hypertriglycerideria-induced pancreatitis: a case-