BLUNT ABDOMINAL TRAUMA AND DUODENUM RUPTURE IN A PATIENT WITH SEVERE HAEMOPHILIA B

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SUMMARY:
We present the case of a 4 year old patient with severe haemophilia B, admitted as an emergency case in the Pediatric Surgery and Orthopaedics Clinic Timisoara, by transfer from the county hospital Târgu-Mureș, with a politrauma after a domestic accident. The onset was acute, after the patient stumbled and hit himself on the edge of a metallic object. On admission he presented a worse general status, pale tegument, dry mucous membranes, a temperature of 37.8 ºC, weak pulse of 120 b/min, blood pressure of 86/40 mm Hg. Abdominal examination showed a slightly distended abdomen, which did not move with respiration and presented tenderness, guarding in the epigastric area and right upper quadrant and signs of peritoneal irritation.

After a short period in which the patient was stabilised and paraclinical investigations were conducted, which determined hypovolemic shock and raised the suspicion of massive hemoperitoneum, a surgical intervention was performed. Intraoperative findings were a wide post-traumatic incomplete rupture of duodenum IV with massive hemoperitoneum, extensive duodeno-jejunal intramural haematoma, pancreatic contusion. A intestinal resection of the IVth part of the duodenum and of the first jejunal loop was performed, followed by a termino-terminal duodeno-jejunal anastomosis, drainage of the the hemoperitoneum and peritoneal drainage. Substitution with FIX concentrate was initiated, initially as a continuous intravenous infusion, afterwards in bolus injections. Local and general evolution was favorable, without complications and the patient was released from the hospital on the 12th postoperative day.

Key Words: haemophilia B, politrauma, duodenal rupture, intestinal resection

TRAUMATISM ABDOMINAL CU RUPTURĂ DE DUODEN ÎNTR-UN CAZ DE HEMOFILIE B FORMĂ SEVERĂ

Rezumat:
Se prezintă cazul unui pacient în vârstă de 4 ani, cunoscut cu hemofilie B formă severă, internat de urgență în Clinica de Chirurgie și Ortopedie Pediatrică Timișoara, prin transfer de la Spitalul Județean Târgu-Mureș, cu un politraumatism prin accident casnic. Debutul a fost brusc, în urmă cu aproximativ 24 de ore, când pacientul s-a împiedicat, a căzut și s-a lovit de marginea unui obiect metalic. La internare a prezentat o stare generală influențată, cu tegumente palide, mucoase uscate, temperatura 37,8 ºC, puls filiform,120 bătăi/min, TA = 86/40 mm Hg. La examenul local s-a constatat: Abdomen moderat destins de volum, nu participă la mișcările respiratorii, dureres spontană și la palpare în epigastru și hipochondrul drept, cu contractură musculară generalizată și semne de iritatie peritoneală.

După o scurtă rechilibrare a pacientului și efectuarea investigațiilor paraclinice, care au decelat șoc hipovolemic și au ridicat suspiciunea unui hemoperitoneu masiv, s-a intervenit chirurgical și s-a decelat intraoperator o ruptură post-traumatică incompletă întinsă la nivelul duodenului IV cu hemoperitoneu masiv, hematom masiv difuz duodeno-jejunului, hematom mezenteric, contuzie pancreatică. S-a practicat enterectomie segmentară cu resecția porțiunii IV a duodenului și a primei anse jejunale și duodeno-jejunoanastomoză termino-terminală, evacuarea hemoperitoneului și drenaj peritoneal. S-a inițiat substituție cu concentrat de FIX, inițial în perfuzie endovenoasă continuuă, ulterior în bolus. Evoluția locală și generală a pacientului a fost favorabilă, fără aparția complicațiilor și externare în ziua 12 postoperator.

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CASE REPORT

A 4 year old patient with severe haemophilia B was admitted as an emergency in the Pediatric Surgery and Orthopaedics Clinic Timisoara, by transfer from the county hospital Târgu-Mureș, with a politrauma after a domestic accident. The onset was acute, after the patient stumbled and hit himself on the edge of a metallic object. Approximately 6 hours after the incident, because of the fact that the child presented progressive epigastric and right hypochondrial pain, nausea, bilious vomiting, constipation and failure to pass flatus, the parents took him to the hospital in Odorhei Secuiesc, from where he was redirected to the emergency county hospital Targu-Mures. Because the hospital in Targu-Mures did not have specific substitution for Haemophilia B and because there was a suspicion of acute abdomen, the child was transferred to Timisoara.

On admission he presented a worse general status, pale tegument, dry mucous membranes, a temperature of 37.8 ºC, weak pulse of 120 b/min, blood pressure of 86/40 mm Hg and a frontal haematoma. Abdominal examination showed a slightly distended abdomen, which did not move with respiration and presented tenderness, guarding in the epigastric area and right upper quadrant and signs of peritoneal irritation.

An erect abdominal X-ray on day 1 showed a single air-fluid level in a dilated stomach and a gasless lower abdomen. There was also a suspicion of a lack of continuity of the duodenal wall with duodenal edema/haematoma. Thoracic X-ray showed a slightly dilated pulmonary hilum with a slightly enlarged heart (Fig.1). The head X-ray was normal.

The emergency abdominal ultrasound showed a well defined hyper echoic mass measuring 58x48x56 mm adjacent to the gall bladder and to the right of the lower part of the stomach. The liver, spleen, pancreas and kidneys appeared normal. There was also a diffuse hyper echoic mass in the Douglas space.

Other paraclinical findings were a hemoglobin level of 6 g%, with only 2.110.000 erythrocytes/mm3, raised leucocytes (12400/mm3), low level of trombocytes (78.000/mm3), raised amylasemia, hypoproteinemia, normal prothrombin time(PT) and prolonged partial thromboplastin time(PTT) of 64 seconds(control 30 seconds), hyponatremia, CRP level of 18.4 mg/l, normal transaminases level.

The child received intravenous rehydration and transfusion with 1 unit of whole blood and 1 unit of fresh frozen plasma. Specific substitution with FIX concentrates was started immediately, initially with a a intravenous bolus of 70 UI/Kg and afterwards as a as a continuous intravenous infusion with 4 UI/kg/h. Nasogastric decompression and intravenous antibiotic therapy with Tienam was also started.

Because of the fact that the general status worsened and the child developed progressive signs of peritonism and abdominal distension and also because of the fact that he was hemodinamically unstable, a surgical intervention was carried out.

A median laparatomy was performed. After entering the peritoneal cavity a massive hemoperitoneum of approximately 500 ml was evacuated. The intraoperative findings were a wide post-traumatic incomplete rupture of duodenum IV, continued to the jejunum over a length of approximately 20 cm, extensive duodenal intramural haematoma, pancreatic body contusion and hematoma into the root of the mesentery.

After the American Association for the Surgery of Trauma (AAST) classification system the duodenal trauma was classified grade I (Table 1) and the pancreatic lesion as grade I (Table 2).

A intestinal resection of the fourth part of the duodenum and of the first jejunal loop was performed, followed by a termino-terminal duodeno-jejunal anastomosis, drainage of the the hemoperitoneum and peritoneal drainage.

Fig.1 Patient for years: thoracic X-ray
Substitution with FIX concentrates was given as continuous intravenous infusion according to the recommendations of the World Federation of Haemophilia, initially at a level of 4 UI/kg/h for the first 24 hours, afterwards 3UI/kg/h and then 2,5UI/kg/h from the 2nd postoperative day until the 8th postoperative day (Table 3), when again bolus injections were started at a dosage of 20 UI/Kg/dosis at 12 hours interval until the 12th postoperative day. Postoperative the patient received a unit of erythrocyte mass concentrate. Antibiotherapy was continued for 10 days.

Local and general evolution was favorable, without complications and the patient was discharged from the hospital on the 12th postoperative day on a normal diet, with no abdominal pain, tenderness or masses and normal bowel habit. He has remained well at follow up.

**DISCUSSION**

Blunt pancreatic and duodenal trauma is uncommon, amounting to less than 2% of all abdominal injuries. These injuries often occur during traffic accidents as a result of the direct impact on the upper abdomen of the steering wheel or the handlebars.

The duodenum and pancreas can be injured simultaneously; isolated injuries are rare (<30%). Coexisting injuries are common (50%–98%), with an average of three to four for each patient. Identification of a blunt injury of the duodenum and pancreas may be difficult because imaging findings are often subtle. Delays in diagnosis, incorrect classification of the injury, or delays in treatment can increase the morbidity and mortality considerably.(1,2,3,4)The morbidity and mortality associated with a trauma to the duodenum and

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**Table 1. Scoring Duodenal Injury**

<table>
<thead>
<tr>
<th>GRADE</th>
<th>INJURY</th>
<th>DESCRIPTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Hematoma, laceration</td>
<td>Involvement of a single portion of the duodenum</td>
</tr>
<tr>
<td>II</td>
<td>Hematoma, laceration</td>
<td>Involvement of more than one portion, disruption of &lt;50% of the circumference</td>
</tr>
<tr>
<td>III</td>
<td>Laceration</td>
<td>Disruption of 50%–75% of the circumference of D2; disruption of 50%–100% of the circumference of D1, D3, and D4</td>
</tr>
<tr>
<td>IV</td>
<td>Laceration</td>
<td>Disruption of &gt;75% of the circumference of D2 or involvement of the ampulla or distal common bile duct</td>
</tr>
<tr>
<td>V</td>
<td>Laceration, vascular injury</td>
<td>Massive disruption of the duodenopancreatic complex or devascularization of the duodenum</td>
</tr>
</tbody>
</table>

Note.—Major variables are involvement of one or more parts of the organ, type of injury (hematoma, laceration, or disruption), and involvement of the ampulla and bile duct. The duodenum is divided into the duodenal bulb (D1), descending part (D2), transverse part (D3), and ascending part (D4).

**Table 2. Scoring Pancreatic Injury**

<table>
<thead>
<tr>
<th>GRADE</th>
<th>INJURY</th>
<th>DESCRIPTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Hematoma</td>
<td>Minor contusion without duct injury</td>
</tr>
<tr>
<td></td>
<td>Laceration</td>
<td>Superficial laceration without duct injury</td>
</tr>
<tr>
<td>II</td>
<td>Hematoma</td>
<td>Major contusion without duct injury</td>
</tr>
<tr>
<td></td>
<td>Laceration</td>
<td>Major laceration without duct injury or tissue loss</td>
</tr>
<tr>
<td>III</td>
<td>Laceration</td>
<td>Distal transection or parenchymal injury with duct injury</td>
</tr>
<tr>
<td>IV</td>
<td>Laceration</td>
<td>Proximal transection or parenchymal injury involving the ampulla or bile duct</td>
</tr>
<tr>
<td>V</td>
<td>Disruption</td>
<td>Massive disruption of the pancreatic head</td>
</tr>
</tbody>
</table>

Note.—Major variables are site (proximal vs distal), type of injury (hematoma, laceration, or transection), and state of the main pancreatic duct. The AAST system differentiates between hematomas, contusions, lacerations with and without ductal involvement, and complete organ disruption and considers the site of the injuries.
pancreas are remarkably high. Mortality for pancreatic injuries ranges from 9% to 34%; for duodenal injuries it ranges from 6% to 29%. However, only 5% of the pancreatic injuries and 30% of the duodenal injuries are directly related to the fatal outcome. The variability in morbidity and mortality is caused by several factors: the presence of coexisting injuries, the mechanism of injury, the time to diagnosis, the presence or absence of major ductal injury, and duodenal perforation, which are considered to be predictors of outcome.(5) The probability of complications after duodenal or pancreatic trauma ranges between 30% and 60% and in many cases is the result of missed findings or diagnostic delays or both. Delayed diagnoses and therapeutic interventions often result in a difficult clinical course with a dubious outcome. However, within the first 48 hours after pancreatic injury, most patients succumb to hemorrhage from splenic, hepatic, or vascular injuries.(3,5,6) Organ injuries most commonly associated with pancreatic trauma are hepatic (46.8% of cases), gastric (42.3%), major vascular (41.3%), splenic (28.0%), renal (23.4%), and duodenal (19.3%). The high energy transfer and the proximity of the duodenum and pancreas to other vital structures result in few isolated injuries.(3,5,6) Coexisting injuries and fatal hemorrhage are responsible for early deaths, while infections and multiorgan failure cause most late ones. Approximately one-third of the patients who survive the first 48 hours develop complications related to their pancreatic or duodenal injury. Common complications of duodenal and pancreatic injuries include pancreatitis, pseudocysts, fistulas, intraabdominal abscesses, pneumonia, and anastomotic breakdown, and these are related to the development of multiorgan failure and septicemia.7,8, About 37% of late deaths are primarily attributable to the injury itself and usually occur within 1–3 weeks of the injury or later.7,8,9 The time between the injury and the diagnosis and definitive treatment is an important factor in the development of complications and their resulting mortality. When a definitive diagnosis is delayed for more than 24 hours, up to 40% of patients are at risk of death, as opposed to 11% of those patients operated on within 24 hours. Another study has confirmed these observations and notes that all of the deaths directly related to duodenal or pancreatic injuries occurred in cases in which diagnosis was delayed.(10,11,12) Today, computed tomography (CT) provides the safest and most comprehensive means of diagnosis of duodenal and pancreatic injury in hemodynamically stable patients.(13,14,15,16,17,18) In general, their retroperitoneal location usually protects the pancreas and duodenum from many instances of minor abdominal trauma. Pancreatic and duodenal injuries usually result only from severe anteroposterior compression trauma against the spinal column, mostly in connection with seat belt injuries, deceleration trauma, and handlebar compression trauma. Less common mechanisms of injury are sports injuries, falls, and blows to the upper abdomen. Most blunt pancreatic injuries (>65%) occur in the pancreatic body. Those to the pancreatic tail and head are less common. Blunt pancreatic trauma is more common among children because of more intense transmission of energy and less protection by a usually thinner layer of peripancreatic fat. Force exerted on the right upper quadrant can affect the pancreatic head or uncinate process and cause injuries in the descending and transverse portions of the duodenum. Coexisting injuries can affect the liver, bile duct, gallbladder, right kidney, and ascending colon. Force exerted on the left upper quadrant results mainly in injuries left of the superior mesenteric artery, to the pancreatic body or tail as well as to the transverse and ascending portions of the duodenum. Coexisting injuries can affect the spleen, stomach, and left kidney.(1,3,20,21) Clinical symptoms comprise a triad of leukocytosis, raised serum amylase activity, which can be absent in the first few days, and upper abdominal pain. However, clinical signs are often vague and nonspecific, sometimes even nonexistent. Therefore,

<table>
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<tr>
<th>Hemophilia B</th>
<th>Dosage</th>
<th>Day</th>
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<tbody>
<tr>
<td>Preoperative</td>
<td>40 IU/Kg</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Bolus</td>
<td></td>
</tr>
<tr>
<td>Postoperative</td>
<td>3-5 IU/Kg/h*</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>2-3 IU/Kg/h*</td>
<td>2-10</td>
</tr>
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</table>

*The diluted bottle of FIX concentrate will be administered during a period of 6-8 hours
major pancreatic injuries can occur with initially minimal epigastric symptoms. Blunt pancreatic and duodenal trauma can occur in patients with multiple trauma, and many will have undergone intubation or received sedatives and therefore cannot provide reliable data for evaluation. To obtain a reliable history in an emergency situation is difficult and in many cases barely feasible. Injuries to the pancreas and duodenum are also likely to be masked by most typical coexisting injuries.\(^{(1,3,15,22)}\)

The most common test is the analysis of serum amylase activity. This can be raised, although in up to 40% of cases it remains normal for 2–48 hours after an injury. Repeated testing is recommended, but results do not indicate the severity of the injury. Instead, continuously increased activity or increasing activity are more reliable and have been used for follow-up studies and to monitor the extent of an established pancreatic injury. In addition, the latter features can indicate the need for intervention or operation. Increased activity in general is not specific, as it may occur with salivary gland, facial, small bowel, or hepatic trauma or if the patient is intoxicated.

Serum lipase activity is also not specific for pancreatic injury. It has been used in children with pancreatic trauma but is not considered cost-effective. Trypsinogen-activating peptide has not been fully evaluated so far, and no reliable data are available.\(^{(15,23,24,25)}\)

CT is an essential means of diagnosing traumatic lesions of the duodenum. Experience has shown that nonperforating duodenal conditions can be missed, as only free air or oral contrast medium extravasation are specific signs of duodenal perforation.\(^{(26,27)}\)

Duodenal injuries are the result of penetrating or blunt trauma. It is essential to distinguish between duodenal contusion, hematoma of the duodenal wall, and perforation and disruption; the latter are indications for surgical treatment. If the duodenal injury is associated with a pancreatic injury, distinguishing individual findings at imaging can be particularly difficult. Perforation is most likely to be detected in the descending and horizontal segments. The differentiation of contusion and bowel wall hematoma from perforation is vital.

Duodenal perforation is suspected if there is a retroperitoneal collection of contrast medium, extraluminal gas, or a lack of continuity of the duodenal wall. Duodenal contusion is suspected with edema or hematoma of the duodenal wall, intramural gas accumulations, and focal duodenal wall thickening (>4 mm) as findings of small bowel injury. Fluid or a hematoma in the retroperitoneum, stranding of retroperitoneal fatty tissue, or pancreatic transaction can be present in both conditions.\(^{(15,17,18,27,28,29,30,31)}\)

A study of 27 children showed that retroperitoneal extraluminal free air or oral contrast material on CT images provided a reliable way to differentiate duodenal perforation from contusions of the duodenal wall.\(^{32}\) In another study, CT showed that extraluminal contrast medium was not as beneficial as the presence of free air in detecting duodenal perforation; extraluminal air on CT images was present in three of five children with perforation, but none had extravasation of contrast material.\(^{(26)}\) Most of the contusions of the duodenal wall and hematomas can be treated conservatively, so treatment can vary, depending on whether it is a contusion or a perforation. Mixed attenuation is a sign of duodenal wall hematoma. However, fluid collections in the anterior pararenal space only and thickening of the duodenal wall have been seen in both perforations and duodenal wall contusions.\(^{(27)}\) At present, no specific data are available about the sensitivity and specificity of multidetector CT in the diagnosis of duodenal injuries. Conventional duodenography showed a 54% sensitivity in the diagnosis of perforation after duodenal trauma if CT findings were not clear; therefore, it cannot be recommended as an adjunct to emergency abdominal CT.\(^{(33)}\)

Correct, early diagnosis of pancreatic injuries is essential for injured patients. Detection of the injury patterns by using CT depends on a reliable and robust technique, particularly timing of an emergency CT study after injury and correct timing of the contrast material bolus, as well as the experience of the radiologist involved.\(^{(15)}\)

In 20%–40% of the cases studied, initial CT findings of patients with pancreatic injuries may be within normal limits in the first 12 hours after the injury; however, much of the published data is still based on the single-detector CT technique. CT diagnosis of pancreatic injuries shows variable sensitivity and specificity because many findings are subtle, absent, or at times slow to develop. The sensitivity and specificity of CT in detecting pancreatic trauma of all grades are reported to be around 80%, and grades of injury tend to be underestimated with CT.\(^{(4,20,23,34,35,36,37)}\) CT findings may be subtle, and sometimes the pancreas may appear normal. The integrity of the pancreatic duct is the most important factor in the decision whether or not to operate. CT is limited in detection of pancreatic injuries when only little peripancreatic fat tissue is present and in detection of
subtle pancreatic duct injuries. Specific signs of pancreatic injuries on CT scans are fractures or lacerations of the pancreas, edema or hematoma of the pancreatic parenchyma, active hemorrhage from the pancreas, and blood collections between the parenchyma and the splenic vein.

The patient in our presentation had no abdominal CT, mainly because he was hemodynamically unstable, once he was admitted in our clinic. On the other hand, more than 90% of all pediatric solid organ trauma can be managed by conservative means, if the patient is hemodynamically stable, and does not require a total of more than 40 ml of blood transfusion/kg body weight in two administrations. Perforation of a cavitary organ requires urgent laparotomy.

Another point of discussion is duodenal hematoma. Intramural haematomas have been reported to occur in any part of the gastrointestinal tract from the esophagus to the colon. They usually present with acute abdomen or gastrointestinal obstruction, but are an unusual cause of either of these presentations. In children, they usually follow accidental or non-accidental abdominal trauma and may be a rare complication of procedures such as small bowel biopsy or of bleeding disorders such as the hemophilias or purpuras. Acute abdomen in a child with hemophilia requires consideration of an intra-abdominal hemorrhagic complications.

A conservative, expectant management approach in the form of nasogastric suction and TPN is appropriate in most patients with intramural bowel hematomas. Resolution can be anticipated to occur in 1-2 weeks, though this may sometimes be delayed for 1-2 months. In a patient with an underlying bleeding problem, early blood component replacement therapy to attain and maintain hemostatic levels of deficient factors is crucial in reducing life-threatening complications from internal hemorrhage. Surgical intervention is reserved for the few patients with complications such as perforation, intussusception or persistent obstruction.

Our patient had a grade III duodenal injury and an extensive duodeno-jejunal haematoma, which cannot be managed by conservative means. The importance of promptly identifying and correcting any underlying lesion in an abdominal trauma in a known haemophiliac is of major importance, because such high-risk hemorrhagic events are a cause of acute morbidity and mortality in haemophilia.

REFERENCES

REFERENCES (CONTINUED)

REFERENCES (CONTINUED)


