April 03, 2013

Dear Editor,

Please find enclosed the edited manuscript in Word format (file name: 1618-review.doc).

Title: Intrathoracic Major Duodenal Papilla with Transhiatal Herniation of the Pancreas and Duodenum
Author: Tarkan Jäger, Daniel Neureiter, Clemens Nawara, Adam Dinnewitzer, Dietmar Öfner, Wolfram Lamadé

Name of Journal: *World Journal of Gastrointestinal Surgery*

ESPS Manuscript NO: 1618

The manuscript has been improved according to the suggestions of reviewers:

1 Format has been updated

2 Revision has been made according to the suggestions of the reviewer:

Reviewer I:

1. Title: It could address the presence of cholestasis as part of the clinical scenario.
   - **Author's action:** The title was changed to:

Intrathoracic Major Duodenal Papilla with Transhiatal Herniation of the Pancreas and Duodenum
A Case Report and Review of the Literature

2. Introduction: The Authors refer to the classification for HH that recognizes four types: Type I or sliding HH and Type II, III and IV that all have a paraesophageal component of the hernia. Regarding type I hernias, they state that are "characterized by an intact phrenoesophageal membrane, lack of a peritoneal sack and sliding of the gastric cardia into the posterior mediastinum". However, sliding HH do have a peritoneal sac; traumatic diaphragmatic hernias are by definition

without a peritoneal sac but in type I HH the sliding portion of the upper stomac is comprised within a peritoeneal sac.

- **Author's action:**

Row 71: A comprehensive classification scheme for HH recognizes four types.

CHANGED: HH are categorized into four types.

Row 72: intact phrenoesophageal membrane, lack of a peritoneal sack and sliding of the gastric cardia into the posterior mediastinum.

CHANGED: intact, circumferential lax phrenoesophageal membrane and widening of the muscular hiatal tunnel{Kahrilas 2008 #13}.

Row 77: Typical for a type IV HH is a large defect in the phrenoesophageal membrane, the existence of a hernial sac with herniation of other abdominal organs (pancreas, spleen, colon, duodenum and small intestine)[3,4].

CHANGED: Typical for a type IV HH is a large defect in the phrenoesophageal membrane with herniation of other abdominal organs (colon, spleen, pancreas and small intestine).

REMOVED from References:

4. **Ellis FH Jr**, Crozier RE, Shea JA. Paraesophageal hiatus hernia. *Arch Surg*. 1986;121(4):416-420 [PMID: 3954587 doi:10.1001/archsurg.1986.01400040052007]

3. Case report: It is unclear why an ERCP was included in the work-up of the patient and the Authors could comment on this point. Apparently, a contrast enhanced CT had established dilated intrahepatic and extrahepatic bile ducts due to compression on the common bile duct at the hiatus. As the Authors pointed out, "endoscopic papillotomy or the insertion of a stent was not necessary" at that time because of normal bilirubin levels. Alternatively, a magnetic resonance cholangiography could have been obtained to investigate further the bile ducts.

- **Author's action:**

Comment: The inclusion of the ERCP in the work-up has certainly to do with the fact that

an intrathoracic presentation of the major duodenal papilla is a rarity without any guidelines or recommendations. We agree that a magnetic resonance cholangiography is an option in this situation.

4. Discussion: The Authors refer to the "general recommendation that all HH type IV should be repaired as soon as possible after the diagnosis despite lack of symptoms". Historically, surgical repair was advocated for the treatment of patients with paraesophageal hernias regardless of whether they had related symptoms. This approach stem from retrospective reports showing 30% to 45% incidence of complications and mortality rates up to 50% among patients left untreated (1,2). More recently, however, several authors have questioned the need for repair in truly asymptomatic patients and this issue is therefore controversial (3). Having said that, in the presented case, although the patient was asymptomatic, there were celar-cut signs of compression on the common bile duct and, subsequently, on the heart that warranted a surgical repair. 1. Skinner DB, Belsey RH: Surgical management of esophageal reflux and hiatus hernia. Long-term results with 1,030 patients. J Thorac Cardiovasc Surg, 1967; 53:33-54. 2. Hill LD, Tobias JA: Paraesophageal Hernia. Arch Surg, 1968; 96:735-34. 3. Stylopoulos N, Gazelle GS, Rattner DW: Paraesophagealhernias: operation or observation? Ann Surg, 2002; 236:492-500.

- **Author's action:**

Comment: Thank you for your very valuable comments. The text was added to the manuscript along with the references.

Row 214 - 230: NEW ARRANGED:

Historically, surgical repair was advocated for the treatment of patients with PEH (type II, III and IV HH) regardless of whether they had related symptoms. This approach stem from retrospective reports showing 30% to 45% incidence of complications and mortality rates up to 50% among patients left untreated{Skinner 1967 #29}{Hill 1968 #30}. More recently, however, several authors have questioned the need for repair in truly asymptomatic patients.{Stylopoulos 2002 #18}.

Although our patient was asymptomatic, there were clear-cut signs (compression of the common bile duct) that warranted a surgical repair. Despite our recommendation of surgical hernia repair soon after the diagnosis, we had to respect the declared intention of the patient for a conservative procedure. The uniqueness of this case also supported this dilemma. So we were forced to wait for surgical repair within an emergency situation complicated by a myocardial infarction and reduced general condition.

Our case illustrates the serious sequelae of a HH type IV that can occur when treated conservatively and therefore we recommend that all HH type IV should be repaired as soon as possible after the diagnosis. Despite of the fatal outcome we were encouraged to publish this case to improve future decision finding processes in similar cases.

ADDED to References:

17 **Skinner DB**, Belsey RH. Surgical management of esophageal reflux and hiatus hernia. Long-term results with 1,030 patients. *J Thorac Cardiovasc Surg* 1967; **53**: 33–54 [PMID: 5333620]

18 **Hill LD**, Tobias JA. Paraesophageal hernia. *Arch Surg* 1968; **96**: 735–744 [PMID: 5647546]

19 **Stylopoulos N**, Gazelle GS, Rattner DW. Paraesophageal hernias: operation or observation? *Ann Surg* 2002; **236**: 492-500 [PMID: 12368678 DOI: 10.1097/01.SLA.0000029000.06861.17]

Reviewer II:

1. The evidence of hiatal hernia affected cardiac activity is too weak. CT could not provide any evidence to show the relationship between coronary obstruction and hiatal hernia. Could author show the angiographic picture of this case? That would be more convincing. Otherwise, just change the title of this case report and neglect the description of cardiac acticity and hiatal hernia.
   - **Author's action:** The title was changed to:

Intrathoracic Major Duodenal Papilla with Transhiatal Herniation of the Pancreas and Duodenum

A Case Report and Review of the Literature

Row 69: with cholestasis and impairment of the cardiac action.

CHANGED IN: with consecutive intra- and extrahepatic cholestasis.

Row 83: **DELETED**: and depletion of cardiac activity.

Row 86: REMOVED: Patients are at risk for myocardial infarction essentially due to a mechanical compression of the heart by hernia content.

Row 147: REMOVED: The exploration of the distal LAD and of the left circumflex artery was impossible due to mechanical obstruction caused by pressure of the hiatal content on the coronary vessels (Figure 1B).

Row 202: head of pancreas in association with mechanical obstruction of the heart, resulting in a myocardial infarction.

CHANGED IN: head of pancreas and an intrathoracic duodenal papilla associated with intra- and extrahepatic cholestasis.

Reviewer Chief editor:

1. Abstract: no less than 128 words
   - **Author's action:** The abstract was rewritten (164 words):

Abstract

Transhiatal herniation of the pancreas is an extremely rare condition. In the published literature we found only eleven cases reported in the period of 1958 to 2011. A coincidental hiatal herniation of the duodenum is described in two cases only.

To our knowledge, we report the first case with a hiatal herniation of the complete duodenum and proximal pancreas presenting an intrathoracic major duodenal papilla with consecutive intrahepatic and extrahepatic cholestasis.

A 72-year-old Caucasian woman was admitted to our department with a hiatal hernia grade IV for further evaluation. According to our recommendation of surgical hernia repair soon after the diagnosis of a transhiatal herniation of the proximal pancreas and entire duodenum, we had to respect the declared intention of the patient for a

conservative procedure. So we were forced to wait for surgical repair within an emergency situation complicated by a myocardial infarction and reduced general condition.

We discuss the therapeutic decision making process and a complete literature review of this rare entity.

2. the next number 14-18 seem not appear in the text, please add
   - **Author's action:**

Row 210: ADDED: Further cases in the published literature are described in table 1[14–17].

Further changes:

Row 12: Author ADDED: Tarkan Jäger, Daniel Neureiter, Clemens Nawara, Adam Dinnewitzer, Dietmar Öfner, Wolfram Lamadé

Row 21: ADDED: **Wolfram Lamadé**, Department of Surgery, Helios Spital, Härlenweg 1, 88662 Überlingen, Germany

Row 27: ADDED: Lamadé W made substantial contributions to conception, design and revised for final approval.

Row 74: New entered: PEH

Row 76: New entered: [3]

Row 80: hiatal changed in transhiatal

Row 85: parameters of cholestasis: **CHANGED IN**: cholestasis parameters.

Row 92: ADDED: Caucasian

Row 120: ENTERED: (Figure 1A, B).

Row 124: ENTERED: (Figure 1A, B).

Row 188: paraesophageal hernia **CHANGED IN**: PEH

Row: 233 CHANGED IN: We thank Prof. Kai Matthes (Department of Medicine, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA, USA) for editorial assistance.

3 References and typesetting were corrected

Thank you again for publishing our manuscript in the *World Journal of Gastroenterology*.

Sincerely yours,

Dr. Tarkan Jäger
Facharzt, Universitätsklinik für Chirurgie
Paracelsus Medizinische Privatuniversität
Landeskliniken Salzburg
Müllnerhauptstr. 48
5020 Salzburg Austria
Tel: +43 (0)662|4482-51086
Fax: +43 (0)662|4482-51008
mailto: ta.jaeger@salk.at
http://www.salk.at/