

# Repair of Type II Paraesophageal Hernia with Nissen Fundoplication in a Patient with Von Willebrand Disease and Spondylolisthesis – a Clinical Case Report

Grzegorz Fibiger<sup>1</sup>, Kinga Gładys<sup>1</sup>, Wojciech Fibiger<sup>2</sup>, Artur Pasternak<sup>1</sup>, Mirosław Szura<sup>3</sup>

<sup>1</sup> Department of Anatomy, Jagiellonian University Medical College, Kraków, Poland

<sup>2</sup> Podhale State Vocational University in Nowy Targ, Nowy Targ, Poland

<sup>3</sup> Department of Surgery, Institute of Physiotherapy, Faculty of Health Science, Jagiellonian University, Krakow, Poland

**Corresponding author:** Wojciech Fibiger, Podhale State Vocational University, Nowy Targ, Poland; Email: fibigerw@mp.pl; Tel.: +48 600 631 536

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## Abstract

Hiatal hernias continue to be fairly common in clinical practice. However, the variety of different symptoms presented by patients may hinder establishing the ultimate diagnosis. Nevertheless, currently, the diagnosis of hiatal hernia can be easily established, based on barium swallow radiography. We would like to present a clinical case report of a patient with complex medical history, including von Willebrand disease, degenerative spinal disease, and chronic sinusitis, who was finally diagnosed with hiatal hernia and treated with a standard laparoscopic Nissen fundoplication. Our case focuses on the significance of comorbidities on patients' symptoms, which sometimes may mislead the therapeutic process.

## Keywords

hiatal hernia, general surgery, paraesophageal hernia, Nissen fundoplication

## INTRODUCTION

Hiatal hernia (HH) is defined as the condition when an abdominal organ, most commonly the stomach, is moved through the esophageal hiatus into the mediastinum and cannot be held up in the abdominal cavity. Consequently, the stomach is constantly pulled up by the esophagus while swallowing or in conditions of increased abdominal pressure. Thus, several conditions which may lead to the development of HH were reported, including obesity<sup>[1]</sup>, pregnancy<sup>[2]</sup>, genetic predispositions<sup>[3]</sup>, collagen diseases<sup>[4]</sup> or even extensive physical training<sup>[5]</sup>.

Clinically, four types of HH can be distinguished based on the topographic relations between gastroesophageal

junction (GEJ) and diaphragm.<sup>[5]</sup> Type I, also called the sliding hernia, is the most common one and refers to the state when only the GEJ is displaced superiorly to the diaphragm, into the mediastinum; conversely, in type II hiatal hernia, the gastric fundus is displaced into the thoracic cavity and the GEJ remains in its anatomical location. Type III hiatal hernia is also called mixed type, due to the fact that it contains elements from both types I and II. Finally, type IV of this condition occurs when other organs such as the colon or the spleen move into the thorax from the abdominal cavity. Types II-IV are also known as the paraesophageal hernias (PEH).

HH is clinically significant due to its close relation to the pathophysiology of gastroesophageal reflux disease

(GERD).<sup>[6]</sup> It is important to highlight that patients with the coexistence of GERD and HH have worse overall prognosis and response to the medical treatment.<sup>[7]</sup> The patients with HH mainly present the typical GERD-related symptoms; however, the range of reported symptoms may vary. Therefore, we would like to introduce a case report of a patient presenting with type II PEH, which was accompanied by unusual symptoms and various comorbidities that made the diagnosis and treatment challenging.

## CASE PRESENTATION

The case involves a 51-year-old obese male who initially presented with type 1 von Willebrand disease (vWD) and a long history of degenerative spinal disease in the form of spondylolisthesis at the L5-S1 level of the lumbar spine with bilateral spondylolysis at L5 and thoracic scoliosis. The patient reported hoarseness, cough, and rhinitis, which would appear after longer episodes of speaking and sitting. The symptoms he originally presented with were thought to be the consequence of chronic sinusitis; however, the patient subsequently started reporting exacerbation of spinal pain in the thoracolumbar region on the left side after long episodes of sitting, which would be relieved by walking. The computed tomography (CT) confirmed the known spinal findings as well as the degeneration of the intervertebral joints. The physical therapy and rehabilitation dismissed the reported ailments temporarily as pain symptoms started to happen, more frequently, and postprandially. The patient reported their localization in the left thoracolumbar segment, with radiation to the thorax and epigastrium. Finally, the pain appeared daily. Additionally, during the ailments, the patient reported the presence of heart palpitations, dyspnea, shallow breath, and dysphagia with belching. The symptoms were severe enough to cause difficulties in sitting and lying as the patient was forced to establish the standing position, keep a shallow breath, or take drugs such as paracetamol, metamizole, and/or drotaverine in order to relieve pain. Initially, the symptoms persisted for a dozen minutes. The physical examination revealed no ab-

normalities. Furthermore, laboratory results were also within the normal ranges. Ultimately, chest X-ray examination was performed and suggested the presence of HH (Fig. 1A). Interestingly, both the prior X-ray examination and previous CT did not detect the mentioned finding. Finally, the X-ray with contrast (Fig. 1B) of the upper part of the digestive tract and the chest CT (Fig. 1C) confirmed the presence of PEH measuring approximately 77×90×59 mm.

Standard endoscopy of the upper part of the digestive tract revealed PEH with intact GEJ and no lesions in the mucosal lining of the esophagus and stomach were found. Subsequently, the patient was consulted with a hematologist for the type 1 vWD with familial occurrence, when the desmopressin test was performed. Desmopressin was administered intravenously in a dose of 0.3 mcg/kg and it provoked a relatively good hemostatic response. After completing the diagnostic evaluation, the patient was scheduled for surgery. In order to provide a hematological protection of the surgical treatment, the use of desmopressin in infusion was recommended. Ultimately, laparoscopic Nissen fundoplication was performed with the suture of diaphragm crura. The procedure went uneventful. The patient was discharged in good condition and reported complete recovery and relief of symptoms during the routine 1-year follow-up.

## DISCUSSION

Paraesophageal hernia is a relatively common disorder with a slight female predominance.<sup>[8]</sup> In terms of etiology, PEH can be either congenital or acquired with a higher prevalence in elderly due to the age-related loss of elasticity of surrounding tissues and muscle weakness of the superior border of the abdomen. The most common obstructive symptoms are dysphagia, postprandial fullness, early satiety, vomiting and/or epigastric pain. The patient may also complain from chest pain or shortness of breath as a result of the compression of structures in the thoracic cavity caused by the expanding stomach. Less frequently reported symptoms are Cameron's ulcers, gastric volvulus,



**Figure 1.** A. The chest X-ray examination in posteroanterior view revealing the presence of hiatal hernia, measuring 95×60 mm, seen as the brightening region which blurred the contour of the diaphragm; B. The chest radiograph with the barium swallow test in the posteroanterior view; C. Computed tomography of the chest in the axial projection revealing the hiatal hernia, measuring 77×90×59 mm with the significant part of the stomach in the thoracic cavity.

and iron-deficiency anemia. Interestingly, the presented patient did not report typical symptoms, as he complained mainly of spinal pain, although, the recent literature suggests that patients with degenerative spinal diseases, such as scoliosis, are at greater risk of developing HH.<sup>[9]</sup> According to SAGES guidelines, the preferred surgical approach for managing HHs is laparoscopic Nissen fundoplication.<sup>[10]</sup> Various literature reviews and studies advocate the superiority of laparoscopic fundoplication, over the open classical counterpart in terms of efficacy, mortality, and cosmetic outcomes.<sup>[11]</sup> Interestingly, a correlation between the increasing size of the HH and a higher risk of recurrence was also revealed.<sup>[12]</sup> Esophageal hiatal hernias have been reported to affect 10% to 50% of the population, increasing with age and with a slight female predilection; therefore, if these patients have upper dyspeptic symptoms, we should suspect them.<sup>[13,14]</sup>

## CONCLUSION

Hiatal hernia in patients with complex medical history may mislead the diagnostic process. Thorough analysis of existing comorbidities' symptoms and additional investigations would enhance proper diagnosis for the right and timely treatment.

## Author contributions

G.F.: conceptualization, writing the original draft; K.G.: writing the original draft; W.F.: writing the original draft; A.P.: conceptualization, writing and editing; M.S.: conceptualization, supervision, writing and editing.

## Conflict of Interest

The authors declare no conflict of interest.

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# Пластика параэзофагеальной грыжи II типа с помощью фундопликации по Nissen у пациента с болезнью Von Willebrand и спондилолистезом – клинический случай

Грегорц Фибигер<sup>1</sup>, Кинга Гладис<sup>1</sup>, Войчех Фибигер<sup>2</sup>, Артур Пастернак<sup>1</sup>, Мирослав Шура<sup>3</sup>

<sup>1</sup> Кафедра анатомии, Медицинский колледж Ягелонского университета, Краков, Польша

<sup>2</sup> Подгальское государственное высшее профессиональное училище в Новы-Тарге, Новы-Тарг, Польша

<sup>3</sup> Кафедра хирургии, Институт физиотерапии, Факультет медицинских наук, Ягелонский университет, Краков, Польша

**Адрес для корреспонденции:** Подгальское государственное высшее профессиональное училище в Новы-Тарге, Новы-Тарг, Польша; E-mail: fibigerw@mp.pl; тел.: +48 600 631 536

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## Резюме

Хиатальные грыжи продолжают довольно часто встречаться в клинической практике. Однако разнообразие различных симптомов, предъявляемых пациентами, может затруднить постановку окончательного диагноза. Тем не менее, в настоящее время диагноз хиатальной грыжи можно легко установить на основании рентгенографии с приёмом бария. Мы хотели бы представить отчёт о клиническом случае пациента со сложной историей болезни, включая болезнь von Willebrand, дегенеративное заболевание позвоночника и хронический синусит, у которого наконец была диагностирована хиатальная грыжа и проведена стандартная лапароскопическая фундопликация по Nissen. В нашем случае основное внимание уделяется значимости сопутствующих заболеваний для симптомов пациентов, которые иногда могут ввести в заблуждение терапевтический процесс.

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## Ключевые слова

хиатальная грыжа, общая хирургия, параэзофагеальная грыжа, фундопликация по Nissen

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