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PRACA POGLĄDOWA
REVIEW

Hepatic portal venous gas in children: why ultrasound matters more than ever – a literature review

Gaz w żyle wrotnej u dzieci: dlaczego ultrasonografia ma dziś większe znaczenie niż kiedykolwiek – przegląd piśmiennictwa

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ABSTRACT

INTRODUCTION: Hepatic portal venous gas (HPVG) is a rare, yet alarming finding in pediatric patients that is historically linked to a mortality rate reaching 75%. However, advancements in imaging techniques now reveal that in many cases it is transient and benign. In this study, we focus on the growing role of ultrasonography in diagnosing pediatric patients with HPVG, highlighting its value in clinical practice.
MATERIAL AND METHODS: A literature review was conducted using PubMed and Google Scholar. The search terms were "hepatic portal venous gas," "pediatrics," "ultrasonography," "diagnostic imaging," "necrotizing enterocolitis," and related variations thereof.

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STATE OF KNOWLEDGE: HPVG can be detected by multiple imaging methods. Interestingly, ultrasonography stands out from the others for its high sensitivity in HPVG detection, lack of ionizing radiation, and real-time results. In pediatric patients, an extended ultrasound exam not only detects HPVG, but also enables clinicians to stratify risk by analyzing gas distribution patterns and other sonographic markers linked to poorer outcomes. Furthermore, ultrasound aids in refining differential diagnoses by tracking the flow of gas through the intricate branches of the portal venous system. Crucially, when HPVG detection via ultrasound is combined with Gordon's criteria, diagnostic accuracy for necrotizing enterocolitis improves significantly, reaching a specificity of 86% and a sensitivity of 90%. This highlights the growing indispensability of ultrasonography in pediatric care.
CONCLUSIONS: Ultrasonography is a powerful, non-invasive tool that enhances HPVG diagnosis and clinical decision--making. Given its advantages, we propose it as an imaging method of choice for the diagnosis of children with HPVG.

KEYWORDS

pediatrics, portal vein, ultrasonography

STRESZCZENIE

WSTĘP: Gaz w żyle wrotnej (*hepatic portal venous gas* – HPVG) jest rzadkim, lecz alarmującym objawem u pacjentów pediatrycznych, historycznie wiążącym się ze śmiertelnością sięgającą 75%. Jednak dzięki postępowi w technikach obrazowania można stwierdzić, że w wielu przypadkach ma on charakter przejściowy i łagodny. W badaniu skupiono się na rosnącej roli ultrasonografii w diagnostyce dzieci z HPVG, podkreślając jej znaczenie w praktyce klinicznej.
MATERIAŁ I METODY: Przeprowadzono przegląd literatury w bazach PubMed i Google Scholar. Użyto haseł: „hepatic portal venous gas", „pediatrics", „ultrasonography", „diagnostic imaging", „necrotizing enterocolitis" oraz ich wariantów.
STAN WIEDZY: HPVG można wykryć za pomocą różnych metod obrazowania. Spośród nich ultrasonografia wyróżnia się wysoką czułością, brakiem promieniowania jonizującego oraz możliwością uzyskania wyników w czasie rzeczywistym. U pacjentów pediatrycznych rozszerzone badanie ultrasonograficzne pozwala nie tylko na wykrycie HPVG, lecz także na ocenę ryzyka poprzez analizę wzoru rozkładu gazu i innych ultrasonograficznych markerów związanych z gorszym rokowaniem. Ponadto ultrasonografia ułatwia zawężenie diagnostyki różnicowej poprzez śledzenie przepływu gazu w skomplikowanych gałęziach układu wrotnego. Co istotne, połączenie wykrywania HPVG za pomocą ultrasonografii z kryteriami Gordona znacząco poprawia dokładność diagnostyczną martwiczego zapalenia jelit, osiągając 86% swoistości i 90% czułości. Podkreśla to rosnące znaczenie ultrasonografii w opiece pediatrycznej.
WNIOSKI: Ultrasonografia jest skutecznym, nieinwazyjnym narzędziem, usprawniającym diagnozowanie HPVG i wspierającym podejmowanie decyzji klinicznych. Ze względu na zalety badania proponujemy je jako metodę obrazowania pierwszego wyboru w diagnostyce dzieci z HPVG.

SŁOWA KLUCZOWE

pediatria, żyła wrotna, ultrasonografia

INTRODUCTION

In pediatric patients, ultrasonography (US) is one of the most commonly used imaging techniques, thanks to its non-invasiveness, low cost, lack of harmful ionizing radiation exposure, and real-time results [1,2]. Therefore, US is usually the first-line imaging method in children, especially for diagnosing abdominal diseases [1].

A rare ultrasonographic finding is hepatic portal venous gas (HPVG), first described in 1955 [3,4,5]. The mechanisms behind the appearance of gas in the portal system remain unclear. The most common theories are as follows:

1. Increased pressure in the intestines causes trapped air to move through the mural capillaries into the portal venous circulation.
2. Damage to the intestinal mucous membrane moves gas produced by microorganisms from the intestinal lumen to the portal system.
3. Bacteria in the abdominal cavity produce gas, bubbles of which pass into the circulation [3,4].

HPVG can be equated with life-threatening conditions such as bowel ischemia, necrotizing enterocolitis, or bowel wall rupture [3,4,6]. Less than 50 years ago, the mortality rate of patients with portal venous gas was as high as 75% [7]. However, due to technological development, the sensitivity of imaging machines has increased, which resulted in more frequent detection of HPVG, which in numerous cases turns out to be temporary and benign [3,4,6,8]. Non-surgical clinical conditions include food allergy, bowel inflammation, early postoperative period after liver transplantation, upper gastrointestinal barium examination, endoscopic procedures, umbilical vein cannulation, and more [3,4,6,9]. This has resulted in the fatality rate of patients with HPVG decreasing to 29–39% [10,11,12].

It is important to emphasize that HPVG is not a disease by itself and that its detection requires an extension of the diagnostic workup. In this study, we

will consider the increasing role of ultrasonography in diagnosing pediatric patients with HPVG.

MATERIAL AND METHODS

We conducted a comprehensive literature review using the databases PubMed and Google Scholar and the search terms "hepatic portal venous gas," "pediatrics," "ultrasonography," "diagnostic imaging," "necrotizing enterocolitis," and all variations related to these terms. The review focused on clinical trials, randomized controlled trials, meta-analyses, systematic reviews, and other review articles. Studies were selected based on their relevance to the topic and quality of evidence.

STATE OF KNOWLEDGE

HPVG imaging methods

Plain radiography

The most basic technique of visualizing gas in the portal venous circulation is plain radiography [4]. HPVG presents as branching peripheral radiolucencies in the liver parenchyma that extend up to 2 cm from the liver capsule [7]. However, the sensitivity of X-ray imaging in detecting HPVG is only 12.5%, since it requires the presence of copious amounts of gas [4]. As a result, the identification of this sign on radiography is correlated with a poor prognosis [4].

Computed tomography

Significantly better results in detecting HPVG can be achieved with computed tomography (CT) [4,12]. It allows for the demonstration of even small amounts of gas, thanks to the "lung window" function [4,8]. On a CT scan, HPVG emerges as tubular areas of decreased density in the liver tissue, branching up to 2 cm from the liver capsule [4,12]. The lumens are mainly visible in the non-dependent left lobe and anterior right lobe [4,13].

Ultrasonography

In US, HPVG manifests as hyperechoic, non--shadowing foci flowing through the portal vein or liver tissue, mainly in the non-dependent part of the liver [3,6,14]. Moreover, the use of Doppler and motion modes further increases the sensitivity of the examination [4,6,14,15]. In Doppler imaging, the hyperechoic gas generates characteristic sharp, bidirectional spikes superimposed on the portal vein wave pattern [6,14]. In motion mode, the hyperechoic gas creates typical linear signals, which imitate a "meteor shower" [14].

Comparison of ultrasonography and computed tomography

Hosokawa et al. [16] compared US and CT by describing 25 pediatric patients with suspected intestinal ischemia who were examined for the presence of HPVG using these two methods within 2 days. Forty percent of the ultrasound examinations revealed gas in the portal system, while only 16% of the CT exams did so. In addition, Chevallier et al. [17] reported on three patients with HPVG detected by US, who underwent abdominal CT within 15 minutes. In each case, the CT was negative for gas in the portal venous circulation, while the subsequent ultrasound examination was able to detect it. These results may indicate a higher sensitivity of US in the detection of HPVG compared to CT, which can be explained by the following factors:

1. the physical basis for gas detection – a small amount of gas can be easily detected by its high impedance on US, while a small volume of gas does not sufficiently alter the density to make it detectable on CT
2. the high blood solubility of the gas – the absence of accumulated gas bubbles prevents their visualization on CT, while US easily shows a small amount of gas
3. the time required for examination – CT is a one--time examination and gas bubbles that are large enough to be detected become untraceable while moving; US is performed over several minutes, which makes it possible to visualize flowing trapped air [17,18].

Computed tomography radiation risk

CT is the method of choice for detecting HPVG in adults, but not pediatric patients [3,4]. The explanation for this is that in children, CT has a significantly higher risk of adverse effects from a given dose of radiation [19]. This is due to their greater radiosensitivity (because of the greater proportion of dividing cells) and longer expected lifetime (during which radiation--induced tumors may develop) [19]. Moreover, due to the thinner torso and smaller cover of organs from the radiation exposure, pediatric patients need lower doses of radiation, which are surprisingly often not reduced [19,20]. As a result, the estimated organ dose after abdominal CT in children is much higher than in adults [19]. The situation is additionally complicated by the fact that CT machines show an absorbed dose of radiation during the scan using the volumetric CT dose index (CTDIvol), which was based on measurements performed on 16-cm or 32-cm phantoms and is independent of the actual patient's size [21]. This is particularly important for neonates, whose bodies are frequently much smaller than even the phantoms.

Strauss and Goske [21] showed that for an abdominal CT scan of an infant with a 6-cm-diameter trunk, the CTDIvol based on a 32-cm phantom indicates 30% of the real absorbed dose; the CTDIvol based on the 16-cm phantom indicates 80% of the real absorbed dose. With this in mind, it is not surprising that, according to research by Brenner et al. [22], the estimated lifetime attribute risk of death from cancer after abdominal CT in a 1-year-old child is estimated at 1 to 550.

Prognostic value of ultrasonography in patients with HPVG

HPVG patterns in ultrasonography

As mentioned before, the mere detection of HPVG does not determine the diagnosis, since it can be the first symptom of either terminal or harmless illnesses. Significantly, extended ultrasound examination after gas is detected in the portal vein system has been shown to help predict the patient's prognosis (Table I).

One study distinguished three patterns of HPVG on US:

1. dot-like pattern – branched distribution of hyperechoic foci throughout the liver; most of the liver tissue is visible, which corresponds to a small amount of gas in the portal venous system
2. streak-like pattern – streaky distribution of hyperechoic gas shadow extending to the peripheral part of the liver, but not reaching the liver capsule, which corresponds to a large amount of gas in the portal venous system
3. fruit-pulp-like pattern – crowded distribution of hyperechoic gas bubbles reaching up to 1 mm from the liver capsule; most of the liver tissue is almost invisible, which corresponds to the massive involvement of the portal venous system [9].

Interestingly, the dot-like pattern is associated with good prognosis and was commonly transitional in nature [9]. However, the streak-like and fruit-pulp-like patterns were correlated with poor prognosis and required aggressive treatment [9]. This can be explained by the fact that gas patterns on ultrasound are closely related to the amount of gas in the portal venous system – the more gas there is, the worse the prognosis [9].

Ultrasonographic risk factors in patients with HPVG

Moreover, precise ultrasound examination in a patient with HPVG can detect other features categorized as risk factors of poor outcome (Table I). Alexander et al. [23] provided an example ultrasound protocol that facilitates the determination of risk of surgery or death. The key components of the examination are an assessment of the bowel wall thickness, echogenicity, dilatation, and peristalsis, as well as the presence of bowel wall pneumatosis, ascites and its type, pneumoperitoneum, or HPVG. The most significant signs are evidence of bowel perforation, such as pneumoperitoneum, focal fluid collection, and complex ascites. In addition, bowel wall thinning, an absence of peristalsis, and perfusion should be taken seriously, as these symptoms may be associated with bowel necrosis, which can lead to perforation. Any of these discoveries requires surgical consultation and, in most cases, surgery due to their association with high mortality. The remaining findings, such as increased bowel perfusion, simple ascites, dilated bowel, HPVG, intestinal pneumatosis, or bowel wall thickening, are not necessarily signs of a serious condition [23].

Table I. Ultrasound results and their impact on prognosis (based on [9,23])

Sign associated with a poor prognosis
Streak-like pattern of HPVG
Fruit-pulp-like pattern of HPVG
Increased bowel wall echogenicity
Bowel wall thinning
Absent bowel peristalsis
Absent bowel wall perfusion
Pneumoperitoneum
Focal fluid collection
Complex ascites
Signs not associated with a poor prognosis
Dot-like pattern of HPVG
Increased bowel perfusion
Simple ascites
Pneumatosis intestinalis
Bowel wall thickening

HPVG – hepatic portal venous gas.

It should be noted that ultrasound alone cannot completely exclude severe disease, but in combination with additional clinical examinations, it facilitates the selection of patients requiring immediate treatment [23]. Furthermore, in case of uncertainty regarding the diagnosis, it enables monitoring of the disease state and the search for the signs classified as risk factors.

Ultrasonography in the diagnosis of patients with HPVG

Identification of HPVG origin by ultrasonography

Ultrasound examination has proven to be a useful diagnostic tool for patients with HPVG. Above all, tracing the gas flow along the branches of the portal vein system with US allows for the determination of the specific parts of the digestive tract from which HPVG originates [24]. For example, gas in the superior mesenteric vein indicates pathology in the small intestine, cecum, ascending colon, or transverse colon; gas in the splenic vein suggests pathology in the spleen or stomach; and gas in the inferior mesenteric vein indicates pathology in the descending or sigmoid colon [24]. This knowledge narrows the differential diagnosis, which increases the likelihood of early,

proper diagnosis and leads to more accurate treatment.

Ultrasonography for necrotizing enterocolitis diagnosis

Until recently, the presence of gas in the portal venous system was considered pathognomonic for necrotizing enterocolitis (NEC), a condition with an overall fatality rate of 23.5% in infants [23,25,26]. This high mortality rate is mainly due to the lack of definitive diagnostic criteria, which frequently leads to delays in diagnosis [23,26,27]. Currently, the most commonly used classification systems are Bell staging and a modified version based on the clinical picture and X-ray findings [27,28,29,30]. However, research shows that the specificity of the modified Bell staging in NEC diagnosis is only 11% [31]. Therefore, it is no surprise that this method is associated with numerous limitations [23,27,32]. Much better results in NEC diagnosis can be achieved using Gordon's criteria (Tables II and III) and ultrasound examination [31,32,33]. Gordon's classification has the main advantage of distinguishing acquired neonatal intestinal diseases in infants, which have a similar clinical course and are often confused [32]. On the other hand, ultrasound examination, which is a sensitive test for detecting HPVG, has satisfactory specificity in NEC diagnosis [4,31,33]. Importantly, Dördelmann et al. [33] reported a specificity of 86% and a sensitivity of 90% for NEC diagnosis by combining ultrasound HPVG detection with Gordon's criteria. It can therefore be assumed that the widespread use of these combined diagnostic methods may contribute to earlier diagnosis of NEC, potentially reducing its mortality and the risk of long-term complications, such as short bowel syndrome or neurodevelopment impairment [25].

Table II. Gordon's criteria for the diagnosis of acquired neonatal intestinal diseases in infants weighing < 1250 grams (based on [32])

Parameter	Feeding intolerance of prematurity	Viral enteritis	Spontaneous intestinal perforation	Necrotizing enterocolitis
Age	< 2 weeks	> 2 weeks	< 2 weeks	> 2 weeks
Feeds (ml/kg/day)	< 80	> 120	< 40	> 80
Clustering	–	+	–	–
Bloody stools	–	+	–	Uncommon
Coagulopathy	–	Related to severity	–	Related to severity
Pneumatosis	–	Common	–	+
Pneumoperitoneum	–	30–40%	100%	20–30%
Ileus	–	Not initially	Variable	+
Surgical finding	None	Ascites, bowel necrosis, distal pneumatosis	Focal perforation of ileum or jejunum	Pneumatosis, mural necrosis
Histological finding	None	Mucosal obliteration, necrosis, inflammation, edema	Robust mucosa, focal necrosis at site of perforation, less inflammation	Mucosal obliteration, necrosis, inflammation, edema
Pathogens	None	Rotavirus, enterovirus	No spec. pathogen	Enterobacteria

Table III. Gordon's criteria for the diagnosis of acquired neonatal intestinal diseases in infants weighing > 1250 grams (based on [32])

Parameter	Cow's milk protein allergy	Viral enteritis	Spontaneous intestinal perforation	Necrotizing enterocolitis
1	2	3	4	5
Age	> 6 weeks	> 2 weeks	< 1 week	< 1 month
Feeds (ml/kg/day)	Cow's milk product	> 120	Unrelated	> 80
Clustering	–	+	–	–
Bloody stools	+	+	–	+
Coagulopathy	–	Related to severity	–	Related to severity
Pneumatosis	Possible	Possible	–	+
Pneumoperitoneum	Rare	Uncommon	+	Less common
Ileus	–	–	Occasional	+

cd. tab. III

1	2	3	4	5
Surgical finding	Ascites, bowel necrosis, distal pneumatosis	Ascites, bowel necrosis, distal pneumatosis	Focal perforation of ileum or jejunum	Pneumatosis, mural necrosis
Histological finding	Mucosal obliteration, eosinophilic inflammation, edema, necrosis	Mucosal obliteration, necrosis, inflammation, edema	Robust mucosa, focal necrosis at site of perforation, less inflammation	Mucosal obliteration, necrosis, inflammation, edema
Pathogens	None	Rotavirus, enterovirus	No spec. pathogen	No spec. pathogen

Limitations of ultrasonography

It should be remembered that despite the numerous advantages of using ultrasonography in pediatric patients with HPVG, this method also has numerous limitations. The result of an ultrasound examination is influenced by multiple variables, such as the patient's condition or the clinician's experience. The barriers to examination on the patient's part include excessive intestinal gas, obesity, and – particularly in children – a lack of cooperation in holding their breath or crying during the examination [1,14,15]. The rarity of HPVG in patients is also significant, as it can result in less training of clinicians in the detection of this symptom, which limits its detection [3].

Another problem is that there is a group of diseases that resemble HPVG on US examination [14]. One condition frequently confused with HPVG is pneumobilia, which also presents as intrahepatic hyperechogenicity [14,34,35]. However, to make the correct diagnosis, it is crucial to be aware that for HPVG, because of the blood flow direction in the portal vein, gas mainly distributes in the peripheral part of the liver, while for pneumobilia, because of the bile flow direction, gas tends to accumulate centrally, further than 2 cm from the liver capsule [6,15]. Other diseases that should be distinguished from HPVG include air in the intrahepatic inferior vena cava and liver abscess [14]. It is important to underscore that all of these conditions require different types of treatment. Therefore, the physicians performing ultrasound examinations must be very familiar with the principles of differentiating between these diseases.

CONCLUSIONS

HPVG is a rare discovery that may be associated with many conditions and does not necessarily indicate a serious illness. We propose US as an imaging method of choice in diagnosing pediatric patients with HPVG, as it is highly sensitive to gas detection, enables the selection of patients with poor prognosis, and helps with diagnosis. We believe that the wider use of US will improve the treatment of pediatric patients with abdominal diseases.

Authors' contribution

Study design – K. Ceglarz, S. Pucyło

Data collection – K. Ceglarz, S. Pucyło, M. Skweres, J. Pielaciński

Manuscript preparation – K. Ceglarz, S. Pucyło, M. Nieczyporuk

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