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Dysphagia as an interdisciplinary problem. Difficulties in the diagnostic process of the vascular anomaly: right-sided aortic arch with aberrant left subclavia artery (case report)

Dysfagia jako problem interdyscyplinarny. Trudności diagnostyczne dysfagii w przebiegu wrodzonej wady naczyniowej pod postacią prawostronnego łuku aorty z lewostronną atreria lusoria (opis przypadku)

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Manuscript Preparation
E – Literature Search
F – Funds Collection

Barbara Maciejewska[1ABCDEFG], Karolina Kania[2ABCDEFG], Piotr Kowal[2ABCDG], Bożena Wiskirska-Woźnica[1ABCDG]

[1]Department and Clinic of Audiology and Phoniatry, Poznan University of Medical Sciences, Poznań, Poland
[2]Clinic of Neurology, Poznan University of Medical Sciences, Poznań, Poland

Article history: Received: 13.06.2017 Accepted: 15.02.2018 Published: 30.03.2018

ABSTRACT: Dysphagia is an underestimated health problem. At the same time, it is a potentially life-threatening condition. Dysphagia in young adults is rare and thus it is rarely discussed in literature. Vascular anomalies are much less frequent causes of dysphagia than structural pathologies of the digestive system, iatrogenic lesions or neurological causes. This paper presents a case of a 21-year-old woman with escalating dysphagia in the course of congenital vascular anomaly in the form of a right-sided aortic arch with retroesophageal left subclavian artery (left arteria lusoria) and compression. The paper highlights delayed symptomatology of the congenital defect, reasons behind the long-lasting diagnostic process, and the role of the laryngological – phoniatric examination in order to exclude oral and pharyngeal causes. The differential diagnostics and treatment options are discussed thereinafter.

KEYWORDS: dysphagia, vascular anomaly, arteria lusoria, diagnosis

STRESZCZENIE: Dysfagia jest niedocenianym problemem zdrowotnym, jednocześnie będąc stanem potencjalnie zagrażającym życiu. Dysfagia u młodych dorosłych jest rzadka i rzadko opisywana. Wady naczyniowe należą do znacznie rzadszych przyczyn dysfagii niż patologie strukturalne układu pokarmowego, zmiany jatrogenne czy przyczyny neurologiczne. W pracy przedstawiono przypadek 21-letniej kobiety z narastającą dysfagią w przebiegu wrodzonej wady naczyniowej pod postacią prawostronnego łuku aorty z towarzyszącym nieprawidłowym przebiegiem lewej tętnicy podobojczykowej uciskającej na przełyk. W pracy zwrócono uwagę na późną symptomatologię wady wrodzonej, przyczyny długotrwałego procesu diagnostycznego, rolę badania laryngologiczno-foniatrycznego w wykluczeniu przyczyn ustno-gardłowych. Przedstawiono diagnostykę różnicową oraz możliwości postępowania leczniczego.

SŁOWA KLUCZOWE: dysfagia, wada naczyniowa, arteria lusoria, diagnostyka

 DOI: 10.5604/01.3001.0011.6688

Dysphagia is an underestimated health problem. Nevertheless, its prevalence increases due to the longer survival rate of the society, which results in the growing number of population aged over 65. The severe form is a potentially life-threatening condition. As a consequence, patients may develop malnutrition, dehydration, aspiration and aspiration pneumonia, strangulation. Milder forms considerably deteriorate the quality of life and / or increase the risk of complications (regurgitation, nutritional deficiencies). Apart from medical / health consequences, dysphagia causes an array of psychological and social problems, disturbing family and social relationships. The prolonged time of meals with simultaneous reduction in their volume may reach 45 – 60 minutes, and it results in the consumption of too few calories, as well as it disrupts social relationships [1,12,14].

Dysphagia results from the difficulty in ingestion and passage of food from the oral cavity through the pharynx to the stomach in the course of morphological damage of the upper alimentary tract or functional disturbances of the nervous and muscular systems. The difficult passage of the food bolus from the oral cavity into the stomach may be classified as high / oropharyngeal dysphagia – connected with the discoordination of the individual swallowing and breathing processes, and low / oesophageal dysphagia – resulting from the difficulty in moving the food through the oesophagus to the stomach [1, 14].

Swallowing disturbances are a symptom occurring in the course of many illnesses. Swallowing disturbances often are a result of ischaemic strokes of the brain or neurodegenerative diseases (motor neurone disease, Parkinson's disease, Guillan - Barre syndrome, multiple sclerosis, cranial nerves neuropathies) [1,5,12,14]. Due to a multitude of aetiological factors, diagnostics of dysphagia requires an interdisciplinary approach – of a laryngologist / phoniatrician, neurologist, gastroenterologist, or even a cardiologist or psychiatrist.The main causes of dysphagia are presented in Table I.

Dysphagia in young adults is very rare and rarely described. The average age of a patient with dysphagia, according to Hoy, is 62 years of age, and the analysis of 2,511 articles on dysphagia, performed by Roden, revealed that as much as two-thirds of them involved patients over 50 years of age [1]. This paper presents a case of a 21-year-old woman with discrete symptoms of cerebral palsy, with escalating dysphagia in the course of a congenital vascular anomaly in the form of the right aortic arch with accessory / aberrant left subclavian artery, the so-called arteria lusoria. This term refers to the abnormal, retroesophageal course of the left or right subclavian artery [13]. This case is especially interesting with respect to the patient's age, the rarity of the cause of dysphagia, and its relatively delayed symptomatology as well as the delays in diagnosis. The emphasis has been placed on the role of the proper laryngological – phoniatric examination in order to exclude oropharyngeal causes, which was a reason to extend the diagnostic process.

Tab. I. Główne przyczyny dysfagii.

CHOROBY	PRZYCZYNY
Nerwy czaszkowe	Neuropatia (porażenie nerwu twarzowego Cukrzycowa neuropatia nerwu błędnego Zespół Guillaina-Barrégo)
Kora mózgowa i pień mózgu	Choroba naczyniowo-mózgowa Stwardnienie rozsiane Choroba neuronu ruchowego Choroba Parkinsona Choroba Alzheimera Porażenie mózgowe
Połączenie nerwowo-mięśniowe	Miastenia Zespół Eatona-Lamberta Botulizm
Mięśnie	Polyomyositis Dermatomyositis (zapalenie skórno-mięśniowe) Miopatia Dystrofia
Zapalenia	Opryszczka Drożdżyca Zapalenie gardła Radioterapia Zapalenie migdałków podniebiennych Zapalenie żołądka (GERD)
Wady strukturalne	Wrodzony rozszczep podniebienia, Pierścień Schatzkiego, pierścienie naczyniowe Nabyte – uszkodzenie, resekcja
Guzy	Nowotwory złośliwe języka, warg, podniebienia, jamy ustnej, nosowej części gardła Guzy mózgu, krtani, przełyku, śródpiersia

Tabela powstała w oparciu o piśmiennictwo [1, 2, 10, 13].

CASE STUDY

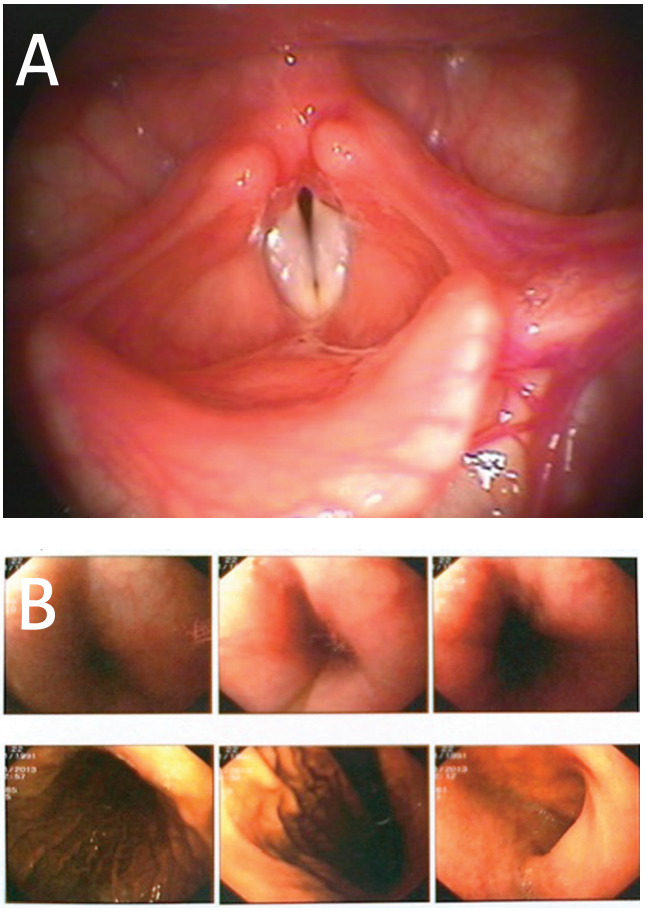
D.S., a 21-year-old woman, with swallowing disturbances escalating for approximately three years, was admitted to the Department of Phoniatrics and Audiology of Poznań University of Medical Sciences (2015). Dysphagia was persistent, during all the meals, disrupting the patient's every-day functioning. It was accompanied by odynophagia, an intermittent pressure sensation in the laryngeal area (more accurately, the symptoms were located on the level of the suprasternal notch). The patient's history included: cerebral palsy (in childhood, muscular hypotonia, at the moment the patient was fully independent, no muscular power deficits). The initial symptoms were: growing aversion to eating, and a progressive loss of body mass. Initially, emotional problems were suspected (the onset of symptoms in the final year of secondary school), and as the loss of body mass progressed as a result of nutritional restric-

tion – anorexia nervosa. In outpatient treatment, the pharmacological treatment was instituted after the psychiatric consultation, with no effect. The verification of the diagnosis was attempted. Gastroscopy, performed in order to exclude the hyperplastic lesions in the lumen, gastroesophageal reflux or oesophageal achalasia, was unremarkable. No abnormalities were detected on the laryngological examination of the larynx. Endocrinological diagnostics was performed for any possible thyroid dysfunctions, which were later excluded (euthyreosis with the proper echogeneity of thyroid parenchyma in ultrasound). MR of the cerebrum revealed only single ischemic foci, apart from that – nothing abnormal detected, including the proper image of the brain stem.

On admission the patient presented with:

- BMI 17.8,
- Slight facial asymmetry and features of a slight left facial paralysis II/III according to House and Brackmann's scale,
- Open nasality (always present in history), velopharyngeal insufficiency,
- Full tongue mobility,
- Two screening tests for dysphagia included water-swallowing test (Daniels 1997) [15] and the Gugging Swallowing Screen (GUSS) for semisolid, liquid, solid swallow trials (Trapl 2007) [16] were correct and did not reveal any remains in the oral cavity (normal oropgaryngeal phase) or any aspitarion,
- ingestion of foods of only small amounts and the prolonged oral phase resulting from the fear of pain / discomfort accompanying swallowing,
- After swallowing, pain localised in the neck and pharynx,
- No cough or choking,
- Uninhibited respiration,
- No food residues in the oral cavity.

On laryngoscopic examination of the larynx, performed several times during hospitalization, the epiglottis was formed properly, vocal folds were smooth and pale, properly mobile on respiration and phonation, no food residues were present in the oral cavity, in the valleculae and the pyriform sinuses (Fig. 1a). Fexible endoscopic evaluation of swallowing (FEES) for studying the physiology and physiopathology of certain stages of swallowing, particularly the pharyngeal stage, was also correct. Another gastroscopy was added to the previous diagnostic procedures, and its findings were as follows: oesophagus without morphological lesions, susceptible to air inflation, its upper sphincter was loose; in the empty stomach, the fluid contents were clear, the mucosa was normal, the inversion – the fundus and the cardia – normal, the pylorus unobstructed,



Ryc. 1. Endoscopic views of the patient's larynx (a – laryngoscopy) and the esophagus (b – gastroscopy).

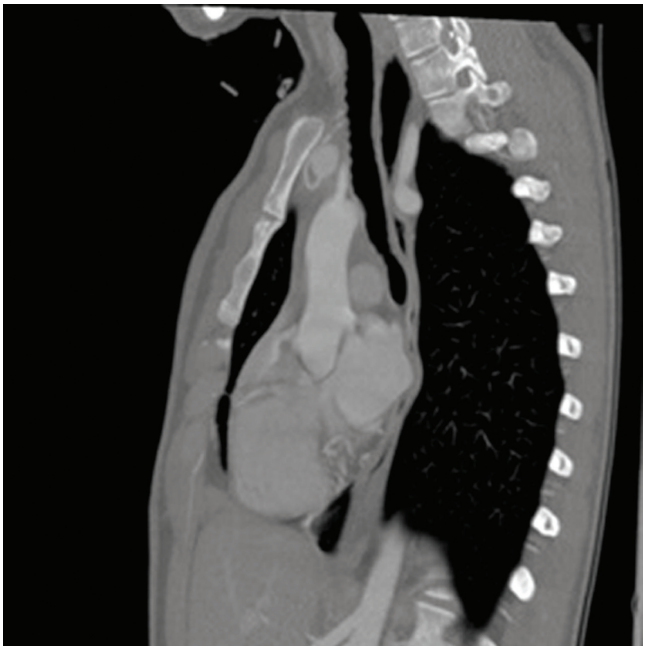
the duodenal bulb and further parts of the duodenum unremarkable, the observation for oesophageal stricture was not confirmed (Fig. 1b).

Due to dysphagia and hypernasal speech, after the neurological confirmation, the patient was diagnosed towards myasthenia – electromyography revealed no conduction problems, the repetitive nerve stimulation test was unremarkable, anti-acetylcholine receptor antibodies (ARAB) were absent. In order to exclude Eagel's syndrome, the patient's skull was x-rayed (the styloid process was within normal limits, 2.8 mm). The contrast examination of the upper part of the alimentary tract with barium meal revealed compression of the vessel on the oesophagus at the level of the aortic arch, which narrowed the oesophageal lumen (Fig. 2). Further diagnostics involved computer tomography of the thorax with contrast (Fig. 3), which proved valuable diagnostically. It revealed the vascular anomaly in the form of the right-sided aortic arch with aberrant left subclavian artery, compressing the oesophagus, with the

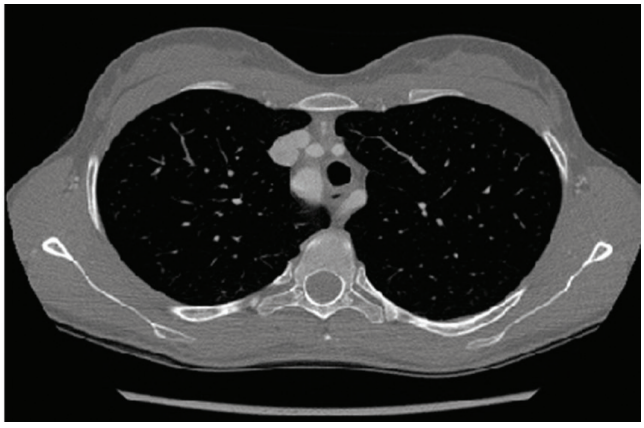
distension of the oesophagus in the proximal part by approximately 2 cm. The patient was referred to the cardiac surgery department, where the detailed vascular reconstruction was performed (angio-MR test, performed with Siemens Avanto apparatus, 1.5 Tesla, with intravenous administration of the contrast medium in TWIST dynamic sequences and delayed VIBE sequences) (Fig. 4, 5), and the surgical treatment was planned. The patient underwent the procedure of endovascular closure of the proximal part of the aberrant left subclavian artery – the vascular malformation was embolized with the use of the vascular plug (t.2, 16 mm in diameter). The follow-up angiography of the aortic arch revealed proper vascular flows in the remaining branches of the arch. At present, the patient does not complain of any further problems.

DISCUSSION

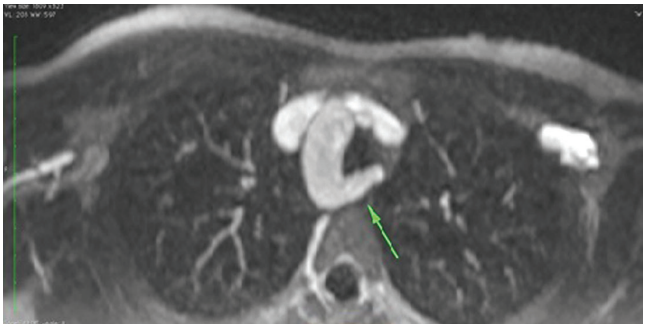
The right-sided aortic arch and the left-sided arteria lusoria are congenital heart defects (0.7 – 1.3%) [13]. Vascular rings may encircle the oesophagus and the trachea partially or completely, thus determining the type of clinical symptoms, such as dyspnoea, stridor, cough, swallowing difficulties, and their intensity. Severe forms are observed most frequently at the early developmental stage – in infancy and childhood, and they are a frequent indication for cardiovascular surgical interventions [11,12,13]. The compression-like symptoms of the trachea and the oesophagus in the form of dysphagia and dyspnoea are often discreet, and symptoms tend to subside as the child grows [13]. Nevertheless, occasionally there occur breathing and / or swallowing difficulties and / or chest pain and / or odynophagia [2,4,17]. The most common symptoms in patients with arteria lusoria originating from RAA are dysphagia, dyspnoea and other respiratory disorders [4]. Symptoms of dysphagia are less persistent than dyspnoea [2,4,9,10,13]. This anomaly is much less frequently diagnosed in older age. Then it becomes a diagnostic challenge. For instance, adult patients, in whom the compression of the trachea dominates, thus resulting in dyspnoea with audible wheezing, are often diagnosed with bronchial asthma [13]. So far, only 32 cases have been published of symptomatic aberrant LSCA in the adult population [4]. The mechanisms that lead to revealing the defect include: the age-related rigidity of the oesophageal wall, age-related elongation of the aortic arch, concomitant aneurysms, the presence of double aortic arch [17]. In the case described above, no additional vascular anomalies were found, thus the first two causes seem the most plausible. Not only age was a factor that delayed the diagnosis. The patient's perinatal history was significant as well. Cerebral palsy, whose symptoms occurred in this case in the form of facial nerve paralysis and palate muscular paralysis, may result in dysphagia due to at



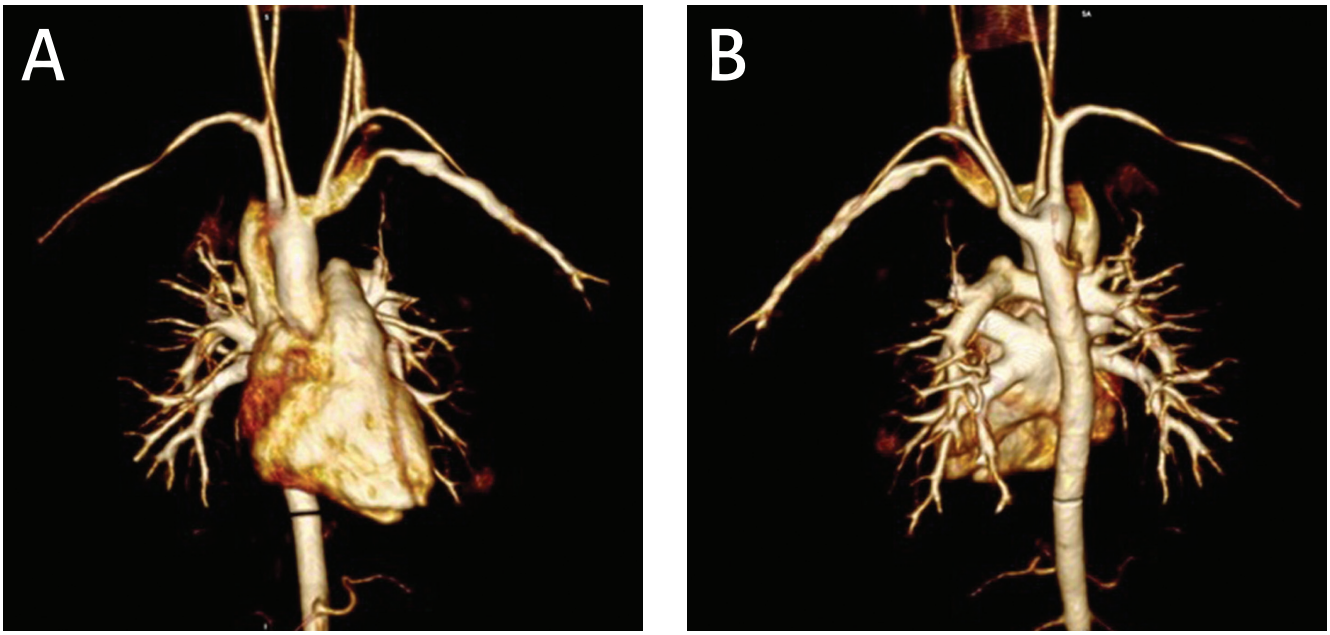
Ryc. 2. Chest X-ray upper part of the esophagus modified barium swallow.



Ryc. 3. CT scan of the thorax showed the right aortic arch with the left subclavian artery with the constricted esophagus..



Ryc. 4. MRI scans of the thorax showed the right aortic arch with the left subclavian artery with the constricted esophagus.



Ryc. 5. 3D reconstructions of the vessels: a) anterior view, b) posterior view

least three factors: weakened tongue motility, delayed onset of the swallowing reflex, and the delayed peristaltic wave of the gastrointestinal tract [1]. Although cerebral palsy in children presents in a variety of types, most cases manifest with swallowing disturbances [7]. Approximately 60% of young adults with cerebral palsy suffers from difficulties with proper processing of food and with excessive accumulation of saliva in the oral cavity, which is often observed with concomitant GERD [6,7].

Clinical and instrumental assessment methods are administrated to find the underlying anatomic and/or physiologic abnormalities leading to swallowing problems and finally to design the appropriate treatment plan. The laryngological and phoniatric examination may contribute to diagnosing dysphagia, especially the oropharyngeal type (the assessment of the structure and mobility of the peripheral speech organs, dysphonia and "moist voice"). The laryngoscopic evaluation is frequently useful during the initial assessment of dysphagia. It allows to observe lesions in the form of food residues, discrete redness of mucosa in the posterior part of the vocal folds and in the interarythenoid notch (indirect symptoms of GERD), as well as enables to diagnose vocal folds palsy and organic lesions localised in the laryngopharynx and the larynx. The absence of abnormalities in the examinations described above should be a reason to search for causes different from laryngological ones, irrespective of the patient's subjective location of the symptoms "somewhere in the voice box". Videofluoroscopy is still the gold standard in dysphagia [3]. The videofluoroscopic swallow study (VFSS) is a commonly accepted "reference standard" instrumental evaluation technique for dysphagia as it provides the most comprehensive information regarding anatomic and physiologic function for swallowing diagnosis and treatment. Flexible endoscopic evaluation of swallowing (FEES) is also available. Fiberoptic endoscopic evaluation of swallowing (FEES) could be a first choice method for studying swallowing disorders on account of the various advantages it offers: easy to use, very well tolerated, allows bedside examination and is economic. Nevertheless, this diagnostic procedure is not without risks, the most probable consequences of which include discomfort, gagging and/or vomiting, vasovagal syncope, epistaxis, mucosal perforation, adverse reactions to topical anaesthetics and laryngospasm [18]. Nevertheless, barium oesophagography is widely accepted to be the gold standard for diagnosing the majority of vascular anomalies [13]. Imaging the aortic arch with the ability to demonstrate not only the arterial branching pattern, but also the relationship of the aorta and its branches to the trachea and bronchi, can be performed by means of the digital subtraction angiography (DSA), computed tomography with contrast, or magnetic resonance imaging (MRI). Once the decision is made to surgically treat the vascular anomaly, many surgeons obtain CT or MR angiogram. The majority of symptomatic patients with arteria lusoria have benefited from surgical intervention such as resection and reconstructive bypass surgery. Janssen et al. concluded that in the absence of another cause of the symptoms and after a trial of medical management, surgery should be considered [12]. Kieffer et al. reported on 19 patients who underwent surgery, 16 of whom had a complete resolution of their symptoms [9].

SUMMARY

1. While conducting differential diagnosis of dysphagia, one should remember that its causes depend on the age group. In neonates and infants, these are neurodevelopmental disturbances and congenital defects; acute infections and traumas are dominant in older children and teenagers; GERD is the most common cause in young adults and middle-aged patients, while the elderly patients are at a considerably higher risk of dysphagia due to neoplastic or neurodegenerative conditions, or their concomitance (according to Altman [1]).
2. The differential diagnosis depends on the location of lesions. Oropharyngeal dysphagia is usually of a neurogenic origin, while the majority of oesophageal dysphagia is related to structural lesions.
3. The anomaly of the right aortic arch with the aberrant left subclavian artery is a rare case of dysphagia, and vascular causes are atypical and non-characteristic for the age group represented by the discussed patient (young adults).
4. The laryngological and phoniatric examination as well as the laryngoscopic assessment may serve as the initial qualification of the level of dysphagia (high / low).

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Word count: 2400 Tables: 1 Figures: 5 References: 18

Access the article online: DOI: 10.5604/01.3001.0011.6688 Table of content: https://otorhinolaryngologypl.com/issue/10687

Corresponding author: Dr n.med. Barbara Maciejewska; Katedra i Klinika Foniatrii i Audiologii, Uniwersytet Medyczny im. K. Marcinkowskiego; 60-355 Poznań, ul. Przybyszewskiego 49; tel.+48 504 772 652; E-mail:barbaramaciejewska@ump.edu.pl

Competing interests: The authors declare that they have no competing interests.

Cite this article as: Maciejewska B., Kania K., Kowal P.,Wiskirska-Woźnica B.: Dysphagia as an interdisciplinary problem. Difficulties in the diagnostic process of the vascular anomaly: right-sided aortic arch with aberrant left subclavia artery (case report); Pol Otorhino Rev 2018; 7(1): 52-58