МРНТИ 76.29.30
https://doi.org/10.35805/BSK2021III028

TYPES OF CORRECTION OF THE SUPRACARDIAC FORM OF PARTIAL ABNORMAL DRAINAGE OF THE PULMONARY VEINS. WARDEN PROCEDURE

Sagatov I.Y.
orcid.org/0000-0002-4668-1513

Dosmailov N.S.
orcid.org/0000-0003-2174-9670

Corresponding author:
Sagatov I.Y. *– Departament head of management of scientific-research working of the JSC "NSCS named after A.N. Syzganov", Almaty, Kazakhstan.*
E-mail: inkar_sagatov@mail.ru

Conflict of interest
The authors declare that they have no conflicts of interest

Keywords
abnormal drainage of the pulmonary veins, Warden procedure, technique of operation

Sagatov I.Y., Dosmailov N.S.
JSC «A.N. Syzganov National Scientific Center for Surgery», Almaty, Kazakhstan

Abstract

The article describes the types of correction of the supracardial form of abnormal drainage of the pulmonary veins. One of the methods of correcting this defect is the Warden operation, which includes: after sternotomy, connection of artificial circulation, cardioplegia, the superior vena cava is cut off, the proximal end is sutured. Next, a right atriotomy is performed, an anastomosis is formed using an autopericardial patch between the abnormal drainage and the left atrium through the ASD. Then an anastomosis is formed between the auricle of the right atrium and the distal end of the superior vena cava. As a result, blood from the abnormal pulmonary veins begins to drain into the left atrium through the ASD.

Өкпе көктамырларының толық емес аномальды дренажының супракардиалық формаларын емдеудің түлері. Warden процедурасы

Хат алысатын автор:
Сағатов І.Е. *– «А.Н. Сызғанов атындағы ҰҒХО» АҚ-ның ғылыми-зерттеу жұмыстарын ұйымдастыру бөлімінің басшысы, Алматы қ., Қазақстан.*
E-mail: inkar_sagatov@mail.ru

Мүдделер қақтығысы
Авторлар мүдделер қақтығысының жоқтығын мәлімдейді

Түйін сөздер
өкпе көктамырларының аномалды дренажы, Warden процедурасы, ота техникасы

Сағатов І.Е., Досмаилов Н.С.
«А.Н. Сызғанов атындағы Ұлттық ғылыми хирургия орталығы» АҚ, Алматы қ., Қазақстан

Аңдатпа

Мақалада өкпе веналарының аномалды дренажының супракардиалды формаларын коррекциялау түрлері сипатталған. Бұл ақауды түзету әдістерінің бірі – Warden отасы, ол мына процестерді қамтиды: стернотомиядан кейін жасанды қан айналымының қосылуы, кардиоплегия, жоғарғы көктамыр қуысы кесіледі, проксималды ұшы тігіледі. Одан кейін оң жақ атриотомия жасалады, жүрекше аралық қалқа ақауы арқылы аномалды дренаж мен сол жақ жүрекше арасындағы аутоперикардты жамау көмегімен анастомоз қалыптасады. Содан кейін оң жақ жүрекшенің құлақшасы мен жоғарғы қуыс көктамырының дистальды ұшы арасында анастомоз пайда болады. Нәтижесінде қан өкпенің аномалды көктамырларынан жүрекше аралық қалқа ақауы арқылы сол жақ жүрекшеге келе бастайды.

Виды коррекции супракардиальной формы частичного аномального дренажа легочных вен. Процедура Warden

Автор для корреспонденции:
Сагатов И.Е. *– руководитель отдела менеджмента научно-исследовательских работ АО «ННЦХ им. А.Н. Сызганова», г. Алматы, Казахстан.*
E-mail: inkar_sagatov@mail.ru

Конфликт интересов
Авторы заявляют об отсутствии конфликта интересов

Ключевые слова
аномальный дренаж легочных вен, процедура Warden, техника операции

Сагатов И.Е., Досмаилов Н.С.
АО «Национальный научный центр хирургии им. А.Н. Сызганова», г. Алматы, Казахстан

Аннотация

В статье описаны виды коррекции супракардиальной формы аномального дренажа легочных вен. Одним из методов коррекции этого порока является операция Warden, которая включает в себя: после стернотомии, подключения искусственного кровообращения, кардиоплегии выполняют отсечение верхней полой вены, проксимальный конец ушивают. Далее проводят правую атриотомию, формируют соустье с помощью заплаты из аутоперикарда между аномальным дренажом и левым предсердием через ДМПП. Затем формируют анастомоз между ушком правого предсердия и дистальным концом верхней полой вены. В результате кровь от аномальных легочных вен начинает дренироваться в левое предсердие через ДМПП.

Introduction

Partial anomalous pulmonary venous drainage (PAPVD) is a rare congenital cardiac defect with the incidence of 0.4%–0.7% and is associated with sinus venosus atrial septal defect (ASD). While most cases are asymptomatic, a patient can present with pulmonary hypertension (PH) and it can be difficult to diagnose. Here, we discuss the case of a young female with PH where thorough investigations lead to a correct diagnosis of PAPVD and ultimately a timely intervention.

PAPDV is an abnormality in which some, but not all, pulmonary veins connect to the right atrium or its tributaries. According to autopsy, the incidence is 0.7% of the population. PAPDV can be combined with other congenital heart defects, most often with an atrial septal defect. Patients with Turner syndrome have a high risk of developing this heart disease.

The first description of vice belongs to Winslow J. (1739). Then, in 1820, Meckel described a case of partial abnormal flow of several pulmonary veins into the superior vena cava. In 1949, Dotter et al made the first report on the diagnosis of cardiac catheterization. The first documented treatment was in 1953, and in 1956 in the Mayo Clinic, for the first time, the correction of PAPDV in the left brachiocephalic vein and other forms was performed.

Partial abnormal pulmonary venous drainage (PAPVD) is an abnormality in which some, but not all, of the pulmonary veins connect to the right atrium or its appendage. According to autopsy data, the incidence is 0.7% of the population. PAPVD can be associated with other congenital heart defects, most often with an atrial septal defect. Patients with Turner syndrome have a high risk of developing this heart defect.

The first description of the defect belongs to Winslow J. (1739). Then, in 1820 Meckel described a case of partial anomalous flow of several pulmonary veins into the superior vena cava. In 1949 Dotter et al. made the first report on cardiac catheterization diagnosis. The first documented treatment belongs to 1953, and in 1956, the Mayo Clinic performed the first PAPVD correction into the left brachiocephalic vein and other forms.

The hemodynamic and clinical manifestations of PAPVD correspond to those in atrial septal defect. The degree of hemodynamic change depends on the number of abnormally connected pulmonary veins. The pulmonary veins can flow into the SVC, into the place of the SVC and brachiocephalic vein fusion above or directly into the azygos vein.

Types of research for PAPVD include: magnetic resonance imaging, CT angiography (the most informative research methods), echocardiography (transthoracic, transesophageal), cardiac catheterization, plain chest X-ray, electrocardiography.

Warden procedure

Clinical case. An 18-year-old female patient was admitted with complaints of general weakness, fatigue, palpitations, recurrent heart pains and frequent colds. CHD was diagnosed in childhood. On echocardiography on admission: right atrium: 4.9x5.4 cm, right ventricle: end diastolic size 4.6 cm, medium pressure 30 mmHg., tricuspid valvae: ring diameter 3.9 cm dilated, regurgitation 1-2. Features: valves are compacted, mobile. V. contracta 0.47 cm. EV LV - 71%. In the atrial septum, there is an upper defect without an upper edge. The two right pulmonary veins drain into the vena cava superior with one collector.

ECG: Sinus rhythm, incomplete blockade of the right branch of the His bundle, right ventricle hypertrophy.

Pulmonary angiography from the trunk of the pulmonary artery. Pulmonary artery trunk tonometry.

When angiocardiography reveals from left atrium: The trunk and branches are moderately expanded. The parenchymal phase of both lungs is uniform - reinforced on both sides. With angiocardiography from the left pulmonary artery: the left collector of the pulmonary veins is drained into the left atrium, there is an intensive discharge of contrasting blood from the left atrium to the right atrium due to ASD. With angiocardiography from the right pulmonary artery: the right collector of the pulmonary veins is drained at the mouth of the superior vena cava on the border with the right atrium.

Operation protocol

Longitudinal sternotomy. The heart is enlarged due to the right sections. The vena cava superior is highlighted and the confluence of the left pulmonary veins below v.azygos is noted. The aorta and vena cava were cannulated. Cardio-pulmonary bypass started. Clamp on the aorta. Farmaco-cold cardioplegia in the aortic root. Opened right atrium. During revision of the interatrial septum, the superior sinus venosus defect is determined. V. Azygos is bandaged, stitched and crossed for vena cava superior mobility.The intersection of the vena cava superior above the level of the superior abnormally inflowing pulmonary vein. The cardial part of the transected vena cava superior was closed with an autopericardial patch, and the main part of the vena cava superior was anastomosed with the right atrial appendage with a 6/0 prolene suture. Further, the collector of abnormally draining pulmonary veins was moved into the left atrial cavity with simultaneous closure of the sinusvenosus defect. The next step was the de Vega tricuspid valvae annuloplasty. The hydraulic test of the tricuspid valve showed satisfactory closing function. Drainage tubes into the right pleural cavity and behind the sternum.

The patient was transferred to the specialized department on the 1st day after the surgery. Drainage tubes were removed on the 2nd day after surgery.

Discharge condition:

As a result of the treatment, the patient's condition improved significantly. Pain in the region of the heart was not observed; physical activity, appetite, general condition returned to normal. Body temperature is normal. The skin and visible mucous membranes are clean, of normal color. Vesicular breathing above the lungs. Breath rate 19 per minute. The heart sounds are clear, the rhythm is correct, there are no noises. Heart rate 96 beats per minute. Blood pressure 110/70 mmHg.

Status localis: The postoperative wound healed by primary intention. The patient was discharged in satisfactory condition on the 21st day after the surgery.

Discussion

First described by Winslow in 1739 [2], PAPVD is a rare congenital cardiac defect which is more common in females with an incidence of 0,4%–0,7% in autopsy series [3]; this may overestimate the clinical significance because most cases are asymptomatic. It is different from total anomalous pulmonary venous drainage in which all or most pulmonary veins drain into the right side of the heart. In PAPVD, usually a single pulmonary vein is anomalous, but there can be some exceptions like in our patient wherein two right-sided pulmonary veins were draining into right atrium. The etiology is unknown, but it represents the persistence of embryonic anastomosis between the systemic and pulmonary vein plexus, resulting in one or more anomalously connecting pulmonary veins. Patients with Turner syndrome, in particular, are at increased risk for PAPVD [4].

The most common ASD associated with PAPVD is sinus venosus type of ASD (80%–90% of cases). In about 10% of cases, the ASD is of secundum type. Normally, each PV contributes an average of 25% of the total pulmonary blood flow; however, when a PV connects anomalously to the RA or SVC, blood is preferentially shunted to this anomalous vein because of the lower RA pressure, compared with LA pressure, producing significant volume overload. This is especially true in the presence of systemic hypertension, mitral valve disease, or left ventricular dysfunction, which increases LA pressures. The clinical evidence may not be apparent until the patient reaches middle age. Some authors have suggested that this defect becomes clinically significant when 50% or more of the pulmonary veins anomalously return.

Adult patient may present with dyspnea and occasionally palpitations. Physical examination findings can reveal signs of the right-sided heart failure. The diagnosis can be confirmed with transesophageal echocardiogram [5], but all the pulmonary veins may not be identified, especially in adults. Cardiovascular magnetic resonance imaging (MRI) is rapidly becoming the procedure of choice to diagnose and characterize congenital heart disease, including PAPVD. MRI also provides additional information including quantitation of heart chamber volumes, ventricular mass, and blood flow through the great vessels, especially when other modalities such as echocardiography yield equivocal findings [6,7,8]. Several techniques used in MRI are particularly useful in the diagnosis of PAPVD; these include cardiac MRI which provides enhanced visualization of the pulmonary vasculature including the anomalous pulmonary veins and phase velocity mapping which directly measure the shunt volume (Qp:Qs) noninvasively. Contrast-enhanced computed tomography scanning is an alternative imaging modality that can help in preoperative planning [9].

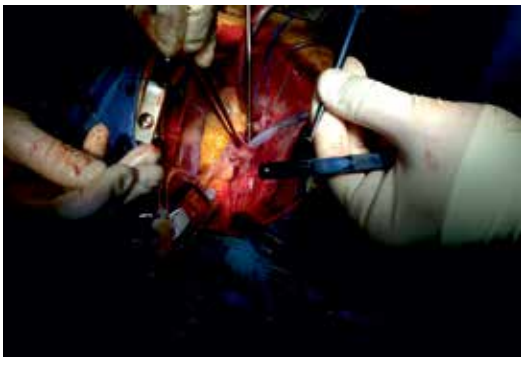

Figure 1. The release of the right atrial appendage trabeculaes

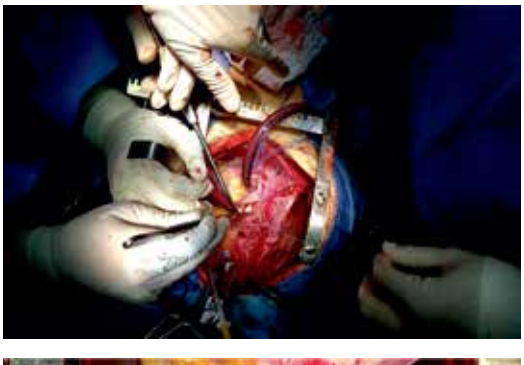

Figure 2. Anastomosis between right atrium appendage and vena cava superior in process

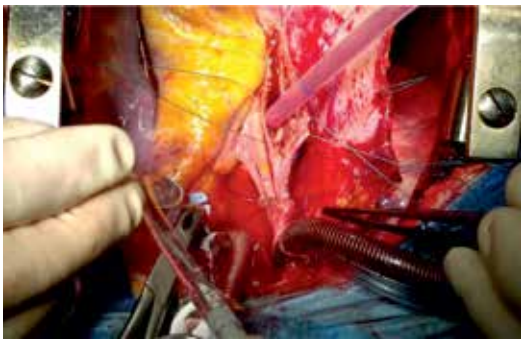

Figure 3. Final look of the anastomosis between right atrium appendage and vena cava superior

Medical therapy is indicated for patients with heart failure or arrhythmias, but the definite treatment is surgical repair, especially when the Qp:Qs is >2,1:1. For the PAPVD to the vena cava superior, the repair techniques may include internal patch technique with or without vena cava superior enlargement, or the caval division technique with atriocaval anastomosis known as Warden technique [10,11]. Patients with internal patch technique must be observed for obstruction of the vena cava superior with vena cava superior syndrome, obstruction of the pulmonary veins, sick sinus syndrome, and supraventricular tachyarrhythmias [12]. The perioperative mortality rate is comparable to that for ASD repair (<0,1%). Prognosis is excellent if surgical repair is done early [13], but it becomes more guarded if the lesion is undetected for a long period.

Conclusion

The advantages of the Warden procedure over the classical operation make it the method of choice for the treatment of the supracardiac form of the partial abnormal drainage of the right pulmonary veins in vena cava superior.

References

1. Ling Y, Gan C, Sandeep B, An Q. Modified Warden procedure for partial anomalous pulmonary venous return to the superior vena cava. J Card Surg. 2016 Oct;31(10):631-634.
2. Yong MS, Griffiths S, Robertson T, Brink J, d'Udekem Y, Brizard C, KonstantinovIE.Outcomes of the Warden procedure for partial anomalous pulmonary venous drainage in children. Interact Cardiovasc Thorac Surg. 2018 Sep 1;27(3):422-426.
3. Sagatov IY., Medeubekov US. Dynamics of urine neutrophil gelatinase-associated lipocalin in cardiac surgery patients in the near term after surgery.Chirurgia 2019 April;32(2):59-61.
4. Sagatov IY., Dosmailov NS., Kvashnin AV., Medeubekov USh., Nurollaeva NA., Ongarbayev KO., Imammyrzaev UY., Saidalin DM. Surgical treatment of the rare congenital heart malformation: quadricuspid aortic valve. The New Armenian Medical Journal Vol.12 (2018), No 3, p. 96-99.
5. Sagatov IY. Modeling of the operational risk in patients with congenital heart diseases. Cardiology, 2015, 131, 29-30.
6. Hofmann M, Dave HH, Schmiady MO, Hьbler M. Warden procedure for correction of sinus venosus atrial septal defect and partial anomalous pulmonary venous connection. Multimed Man Cardiothorac Surg. 2016 Dec 9;2016. doi: 10.1510/mmcts.2016.007.
7. Pavy C, Gavira N, Maminirina P, Baron O. Right partial anomalous pulmonary venous connection to the superior vena cava following the Warden procedure. J Card Surg. 2018 Sep;33(9):565-569.
8. Hopkins KA, Farber MO, Singh R, Kay WA, Herrmann JL, Brown JW. Warden Procedure in a 77-year-old Man. Ann Thorac Surg. 2019 Mar 25. pii: S0003-4975(19)30405-9.
9. Zubritskiy A, Naberukhin Y, Arkhipov A, Gorbatykh Y, Khapaev T, Nichay N, Kulyabin Y, Bogachev-Prokophiev A, Karaskov A. Outcomes of Double-patch and Warden Techniques in Patients with Supracardiac Partial Anomalous Pulmonary Venous Connection. Heart Lung Circ. 2018 Dec 14. pii: S1443-9506(18)32007-9.
10. Aggarwal N, Gadhinglajkar S, Sreedhar R, Dharan BS, Chigurupati K, Babu S. Warden repair for superior sinus venosus atrial septal defect and anomalous pulmonary venous drainage in children: Anesthesia and transesophageal echocardiography perspectives. Ann Card Anaesth. 2016 Apr-Jun;19(2):293-299.
11. Zubritskiy A, Arkhipov A, Khapaev T, Naberukhin Y, Omelchenko A, Gorbatykh Y, Bogachev-Prokophiev A, Karaskov A. The Warden procedure can be successfully performed using minimally invasive cardiac surgery without aortic clamping. Interact Cardiovasc Thorac Surg. 2016 Feb;22(2):225-227.
12. Hiten R. Patel, Suchit Bhutani, Fayez Shamoon, Hartaj Virk. Deciphering a case of pulmonary hypertension in a young female: Partial anomalous pulmonary venous drainage the culprit. Ann Thorac Med. 2018 Jan-Mar; 13(1): 55–58.