CLINICAL CASE OF BIPLASTIC CORRECTION OF ABNORMAL PULMONARY VEIN DRAINAGE WITH SINUS VENOSUS DEFECT IN A DOMINANT INNOMINATE VEIN IN ADOLESCENCE

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

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Abstract

Sinus venosus defectis an unusual type of interatrial communication and is almost always combined with partial abnormal pulmonary vein drainage (PAPVD) into the superior vena cava (SVC) or right atrium (RA). Different types of venous outflow through the vena cava occur: the presence of an additional left vena cava with different drainage loci to both right and left atriaor, as in our case, the dominant unpaired vein directly draining into the right atrium with outprior communication with the superior right vena cava. This paper describes a clinical case of radical correction of partial anomalous pulmonary vein drainage combined with sinus venosus atrial septal defect (ASD) in a 14-year-old child with dominant venous outflow from the upper half of the body via innominate vein, by combined biplasty from auto pericardial patches, showing adequate upper-venous outflow under cardiopulmonary bypass. It is necessary to provide clear information about congenital heart disease and optimal timing of its surgical correction to a wide audience, not only to the professional medical community, but importantly, to the entire population of the Republic. The provision of publicly available professional information should be highlighted on the official websites of clinics and all available media opportunities, where parents and patients could be fully acquainted with the specific soft entire treatment processing an accessible and convenient information platform.

Жасөспірім кезеңдегі басым көлденең венасы бар sinus venosus ақауы бар аномальді өкпе венасының дренажын бипластикалық түзетудің клиникалық жағдайы

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Мүдделер қақтығысы
Авторлар мүдделер қақтығысының жоқтығын мәлімдейді

Түйін сөздер:
өкпе веналарының ішінара ақаулы дренажы, синусты веноздық ақау, клиникалық жағдай, туа біткен жүрек ақауы, жүрекке операция.

Аңдатпа

Sinus venosus ақау – жүрекшеаралық коммуникацияның сирек кездесетін түрі және әрдайым өкпе веналарының ішінара ақаулы дренажымен (ӨВІАД) жоғарғы қуыс венаға (ЖҚВ) немесе оң жақ жүрекшеге (ОЖ) жанама жүретін ақау. Бұл ретте қуыс веналары бойынша веноздық ағудың алуан түрлері бар: оң жақ, сонымен қатар сол жақ жүрекшеге түрлі локустарды дренаждау арқылы қосымша сол жақ ЖҚВ болуы немесе біздің жағдайымыздағы – оң жақ жоғарғы қуыс венамен алдыңғы коммуникациясыз оң жақ жүрекшеге тікелей дренаждалған жұп емес басым венаның болуы. Осы жұмыста 14 жастағы баланың синусты веноздық типті жүрекшеаралық қалқа ақауымен жанама өкпе веналарының ішінара ақаулы дренажын түбегейлі түзету клиникалық жағдайының сипаттамасы ұсынылған. Көлденең вена арқылы дененің жоғары жартысынан вена қанының басым ағуы кезінде, жасанды қан айналымы кезінде адекватты жоғарғы-веналық ағу көрсетілген, аутоперикардиальды жамаудан комбинацияланған биопластика. Жүректің туа біткен ақаулары туралы және оларды хирургиялық түзетудің оңтайлы мерзімдері туралы ақпаратты медицина қызметкерлерінің кең аудиториясына ғана емес, сондай-ақ Республикамыздың бүкіл халқына жеткізу маңызды. Баршаға қолжетімді кәсіби ақпарат клиниканың сайтында және мүмкіндігінше барлық бұқаралық ақпарат құралдарында жарияланууы керек, осы жарияланымдар арқылы ата-аналар және науқастар қолжетімді және қолайлы ақпараттық платформада емдеу процесінің бүкіл ерекшелігімен толық көлемде таныса алуы керек.

Клинический случай бипластиковой коррекции аномального дренажа лёгочных вен с дефектом sinus venosus при доминирующей поперечной вене в подростковом периоде

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Аннотация

Sinus venosus дефект является необычным типом межпредсердной коммуникации и практически всегда сочетается с частичным аномальным дренажем легочных вен (ЧАДЛВ) в верхнюю полую вену (ВПВ) или правое предсердие (ПП). При этом встречаются различные типы венозного оттока по полым венам: наличие дополнительной левой ВПВ с различными локусами дренирования как в правое, так и в левое предсердие или как в нашем случае – наличие доминирующей непарной вены непосредственно дренирующейся в правое предсердие без предварительной коммуникации с верхней правой полой веной. В настоящей работе представлено описание клинического случая радикальной коррекции частичного аномального дренажа лёгочных вен в сочетании с дефектом межпредсердной перегородки (ДМПП) типа sinusvenosus ребёнка 14 лет, при доминирующем оттоке венозной крови от верхней половины тела по поперечной вене, комбинирующей бипластикой из аутоперикардиальных заплат, показавшей адекватный верхне-венозный отток при искусственном кровообращении. Необходимо обеспечить ясную информацию о врождённых пороках сердца и оптимальных сроках их хирургической коррекции широкой аудитории не только профессионального круга медицинских работников, но важно, и всему населению Республики. Предоставление общедоступной профессиональной информации должно быть освещено на официальных сайтах клиник и всех имеющихся возможностей средств массовой информации, где родители и пациенты могли бы в полном объёме, ознакомится со спецификой всего лечебного процесса на доступной и удобной информационной платформе.

Ключевые слова:
частичный аномальный дренаж лёгочных вен, дефект sinusvenosus, клинический случай, врожденный порок сердца, операция на сердце

Introduction

Partial abnormal pulmonary vein drainage (PAPVD) is a congenital heart defect (CHD) in which the pulmonary veins, but not all, flow abnormally into the right venous sections: rightatrium (RA), superior or inferior vena cava, coronary sinus, left unnamed vein. This anomaly was first described by Winslow in1739. PAPVD accounts for less than 1% of all CHDs. The defect is usually combined with atrial septal defect (ASD) and accounts for 14.7% of all atrial septal defects [1-3].Often this anomaly in neglected stagesis accompanied by high pulmonary hypertension, complicating the course of abnormal hemodynamics of the uncorrected malformation [4].

The sinus venosus-type ASD is most commonly located a tor just below the superior vena cava (SVC) junction in the RA. PAPVD is most commonly associated with the valvular sinus ASD. It usually presents as one or more small veins of the upper lobe of the right lung, draining directly into the SVC. Occasionally the interatrial septum is intact [5]. As a gold standard method in topographic diagnosis of abnormal cardiovascular structures of extracardiac location with determination of optimal choiceof surgical correction, computed tomography (CT) techniques are used [6].

Clinical case

Patient O., 14 years old, was admitted to the clinic on June 2, 2022 with the diagnosis: PAPVD. Secondary ASD of sinus venosus type. PFO., with complaints of dyspnea and rapid fatigability on physical exertion. Basic diagnostic tests: ECG, echocardiography (ECHO),contrast computed tomography cardiac angiography were performed. The defect was confirmed by ECHO data: mean right ventricular pressure (MRVP) 35 mmHg., TAPSE: 3.4 cm; interventricular septum: intact; interatrial septum: sinus venosus defect 0.8 x 1.3 cm, PFO 0.3 cm. Interatrial septum in the middle third is thin with small defects of 0.1 cm, 0.1 cm, 0.3 cm. PDA: not visualized. Cardiac cavities: the right parts were dilated. Conclusion: Partial anomalous drainage of the pulmonary veins. ASD. Pulmonary hypertension (PH).

To clarify extracardiac vascular structures, abnormally flowing right pulmonary veins, topographic anatomy of the malformation, and choice of surgical correction method, a CT study was performed (Fig.1): contrast-enhanced Tomohexol 350-100ml I.V. On a series of CT scans, the upper and middle lobe pulmonary veins on the right side flowed into the ERV. The right lower lobe vein and the left upper and lower lobe veins flow into the LA. There is a

communication between the pulmonary veins and the SVC. The aorta at the root is 2.1 cm. Theascending thoracic aorta is 2.3 cm. Aortic arch A segment - 1.5 cm, B segment - 1.3 cm, C segment - 1.4 cm. Descending aorta - 1.6 cm. PA trunk - 2.9 cm, right branch - 1.6 cm, leftbranch - 2.0 cm. The branches of the brachiocephalic trunk, the left common carotid and subclavian arteries are contrasted, unchanged. The superior and inferior vena cava flow into the right atrium. Conclusion: CT picture of a supracardiac form of partial anomalous pulmonary vein drainage.

On 06.06.2022 a planned surgical correction was performed under cardiopulmonary bypass (PB): plasty of PAPVD, sinus venosus ASD with autopericardial patch, suturing of PFO, dilating plasty with autopericardial patch of transverse (innominate) vein. Intraoperatively, the right heartsections were significantly enlarged. During isolation, the right SVC could not be visualized, the innominate vein was dilated and flowed directly into the RA (the place of SVC in flow),the upper right pulmonary veins flowing into the mouth of the SVC were determined from theside. There are cicatrices on the ascending Ao and innominate vein at the level of its horizontal part and the IVC estuary. Aortic cannulation, separate cannulation of the innominate vein,spontaneous hypothermia up to 33 degrees Celsius,beginning of PB. IVC cannulation - complete PB. On revision: sinus venosus ASD 23 mm in diameter, all 4 mouths of the rightpulmonary veins with drainage into both atria (Fig. 2) flow into the mouth of the SVC and the lateral wall of the RA. In the area of the limbus PFO 3 mm. We performed ASD plasty with an autopericardial patch with continuous convolute sutures using Prolene 4/0 thread with transfer of the right PV orifices into the LA cavity. Dilation plasty of the innominate vein andthe orifice of SVC with an autopericardial patch was performed on a working heart. After the clamp was removed from the aorta, cardiac activity recovered on its own, sinus rhythm. Control postoperative ECHO on the 5th postoperative day: Tricuspid valve: 1st degree regurgitation. Mitral valve: 1st degree regurgitation. Aorta: gradient on the descending Ao 9 mmHg. PA trunk: 2.1 cm. LV EF 66%. LV SPD 20 mm Hg. TAPSE: 0.8 cm. Interatrial septum: patch, noshunts. Cardiac cavities: the right side was moderately dilated. In the pericardium there are traces of fluid t/c 0.8 cm. The child with positive dynamics in satisfactory condition was discharged on the 8th postoperative day with recommendations for outpatient cardiac rehabilitation.

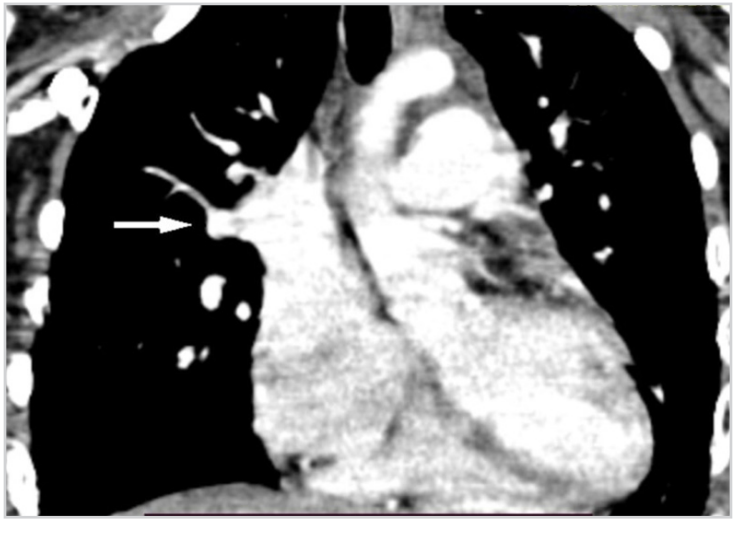

Figure 1.
Frontal CT-section angio-cardiography of the supracardiac form of PAPVD. The arrow shows the in flow of the right pulmonary veins into the SVC

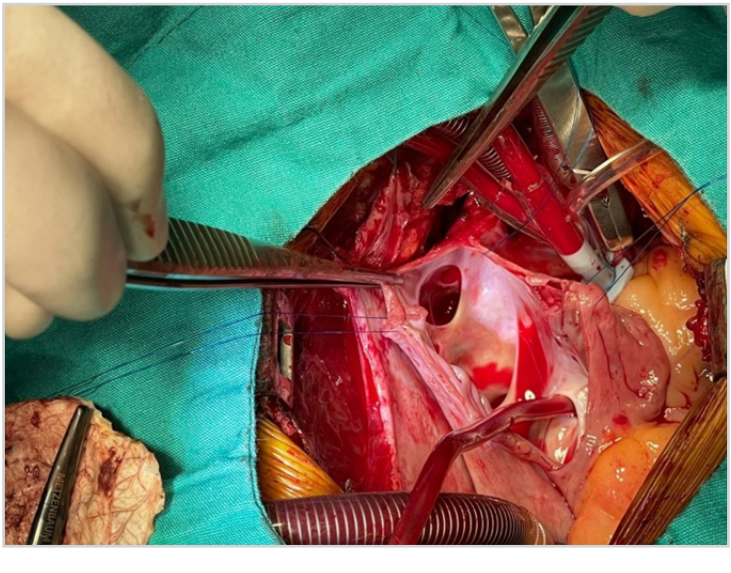

Figure 2.
Intraoperative image of PAPVD. A secondary sinus venosus type ASD is seen, through which abnormally draining right pulmonary veins flowing into the SVC are visualized

Discussion

In later life, these types of malformations are dangerous with the development of high pulmonary hypertension (HPH), complicating their correction. International standards recommend surgery for this type of malformation in infants, asymptomatic patients without PH no later than 4 years of age [7, 8, 9, 10], but the heart defect was detected only this year on ultrasound examination, and they had not sought medical attention before. Within 14 years, the child developed PH.

Intraoperatively, we found unusual architectonics of venous return in the vena cava of theupper body: visualization of the SVC was difficult due to the location of the innominatevein atthe place where it flowed into the RA. The nonspherical vein was larger than usual diameter, had no communication with the SVC and directly flowed into the place where the SVC normally flows. Standard SVC cannulation in partially abnormal right pulmonary drainage begins with placement of a cistern high into the vena cava, but considering the dominance of the unpaired vein, we placed a venous cannula directly into it with the expectation that SVC drainage would go either through the existing collaterals or directly into the RA. The autopericardium patch was used to form a tunnel moving into the LA over the mouths of theright pulmonary veins with simultaneous closure of the ASD. Given the location of the upper part of the tunnel just under the distal incision of the RA and unpaired vein mouth, this area was enlarged with an autopericardial patch.

Conclusion

The highlighted clinical case presented a variant of classical correction of combined PAPVD malformation with sinus venosus type ASD by tunnel biplasty with alternative upper venous cannulation along the dominant unpaired vein, which showed adequate upper venous outflow of the entire artificial cardiopulmonary bypass cycle. It is unacceptable on the part of doctors, as well as parents of children with CHD, to miss the timing of surgical treatment, on which the most favorable outcome of the most complex surgery depends.

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