WEB OF SCIENTIST: INTERNATIONAL SCIENTIFIC RESEARCH JOURNAL
ISSN: 2776-0979, Volume 4, Issue 3, Mar., 2023

ABNORMAL PULMONARY VEIN OCCLUSION (Literature Review)

Mamatov Mukhammadzhon Ahmadovich
Applicant, Republican Specialized Scientific and
Practical Medical Center of Surgery named Academician V. Vakhidov

Abstract:
In this scientific article, the authors describe a review of the literature of total abnormal pulmonary vein drainage (TAPVD). In this regard, the study of the clinical course and methods of diagnosis of TAPVD is relevant, which makes it possible to timely identify the defect, rational planning of the examination and the choice of the optimal method of correction. The definitions, frequency of occurrence, causes of the defect and diagnostic methods are described, including echocardiography, multi-spiral computed tomography with contrast and catheterization of the heart cavities with angio-cardiography. Various types of surgery with different variants of TAPVD are described. Depending on the level of the confluence of the pulmonary veins and the location of the ASD, patients are divided into 4 groups: supracardial, cardiac, infracardial and displaced. The literature review describes the complications of the defect and the results of the operation.

Keywords: congenital heart disease, diagnosis, total abnormal drainage of pulmonary veins, atrial septal defect (ASD), hollow veins, echocardiography, catheterization of heart cavities with angio-cardiography, result.

Introduction

Abnormal pulmonary vein occlusion (abnormal pulmonary vein occlusion) is a complex congenital heart defect in which some (or all) of the pulmonary veins (PVs) flow into the right atrium or its tributaries. Depending on whether all or part of the LV flow into the systemic veins or the right atrium, there are partial and complete abnormal pulmonary vein occlusion. Partial anomalous pulmonary vein occlusion is characterized by one or more, but not all, pulmonary veins flowing into the RV or into the vena cava and its tributaries. The first description of the malformation belongs to J.B.Winslov. Polyal abnormal pulmonary vein inflow is a congenital heart defect, the main feature of which is the inflow of all LV into PP or its tributaries, the communication between the large and small circuits occurs at the atrial level through the ASD. The first description of this malformation belongs to J.Wilson. The incidence of abnormal pulmonary vein occlusion is 0.5-9% of all CHDs. Among all CHDs, total

anomalous pulmonary vein occlusion occurs in no more than 0.7-2.6% of cases. In adolescent and older children, the incidence of total anomalous pulmonary vein occlusion begins to decrease dramatically, and in adults this defect occurs in isolated cases. Some have observed and operated on patients in the 3rd or 4th decade of life. Nevertheless, according to Amosov N.M. et al. Ando H. et al. the absolute majority (80%) of infants die before the age of one year. Such "malignant" course of full abnormal pulmonary vein congestion is associated with early progressive heart failure due to small size of ASD, ICC stasis and severe form of hypoxia. In 38% of cases, complete abnormal pulmonary vein occlusion is combined with other complex heart defects, the presence of which determines the longevity of such patients. Most often, in 60-70% of cases, the defect is combined with ASD or open oval window; rarely - with interventricular septal defect, open arterial duct, etc.

Classification

Numerous classifications of ADLV have been proposed in different years, but none of them has been widely used. Among numerous classifications of abnormal pulmonary vein inflow, the classification proposed by R. Darling et al. is the most widely spread; it is based on anatomical principle depending on the level of LV inflow. The authors distinguish 4 types of malformation: I type - supracardiac or supracardiac: LV enter directly into superior vena cava (ACV) or its tributaries; II type - cardiac or cardiac: LV enter BC or coronary sinus; III type - infracardiac or subcardiac: LVs flow into the inferior vena cava (IVC) or its tributaries below the diaphragm, and IY type - mixed: blood from the LV enters the right heart through two or three pathways described above. These differences in the anatomical structure of the defect determine the specificity of hemodynamic disorders, clinical course, tasks and methods of surgical treatment for each type of defect.

HEMODYNAMICS. Hemodynamics in complete or total anomalous pulmonary vein infiltration is characterized by volume overload of the small circle of circulation (SCC) and depletion of the great circle. In 2/3 of patients there is pulmonary hypertension accompanied by cyanosis and early development of severe decompensation. Pulmonary arterial hypertension in 20% of patients has a hyperkinetic character and in 80% - sclerotic. The presence of pressure gradient between the right and the left atria is of great importance while assessing hemodynamics, it testifies about inadequacy of interatrial defect, the size of which determines the volume of the systemic blood flow. The more blood is supplied to the IAC and there is a pronounced pulmonary-venous obstruction, the faster the volume overload of the right heart and arterial LH develops. At the same time, the left heart retains its normal size or is

hypoplastic. Left ventricular stroke volume and left ventricular minute volume of the great circle of circulation (GVC) are often decreased. Clinic of abnormal pulmonary vein infiltration is varied and depends on many factors. Often the clinical picture of full abnormal pulmonary vein disease is characterized by the development of early and rapidly progressing heart failure, which is the main cause of high mortality of patients, many of whom die before the age of one year. General condition of patients is severe, and only in rare cases - satisfactory. In clinical diagnosis modern methods of investigation are used: electrocardiography; echocardiography; radiography; cardiac catheterization with angiocardiography. There are data in the literature that cardiofibroscopy, MSCT and magnetic resonance imaging have been used in the diagnosis of ADLV. This method gives quite a lot of information about the presence of ASD and abnormal pulmonary vein drainage.

Natural course and prognosis. In the natural course of full abnormal pulmonary vein occlusion with LV the lethality is very high. The average life expectancy of these patients is only 7 weeks, moreover, the presence of an obstruction in the outflow pathway of LV reduces the survival rate to 3 weeks. According to many authors, at least 75% of patients die in the first year of life and only a few patients survive until the age of 3-4 years. In contrast, the natural course of total abnormal pulmonary vein occlusion in patients with normal LA pressure is relatively favorable; the disease proceeds as in a large ASD. There are reports in the literature describing patients survived up to 50-60 years old.

Treatment. In case of full abnormal pulmonary vein congestion, radical surgical treatment is actually the only intervention, after which one can expect a stable positive result. Nevertheless, palliative surgery does not lose its importance, although it can be used only for the treatment of newborns in critical condition - the operation consists of increasing the interatrial communication by balloon arteriseptostomy, thus achieving better conditions for blood flow to the LP, and therefore improving blood circulation in the great circle. However, the operation is of limited effectiveness and is performed in children no older than three months of age. In recent years, other types of palliative interventions have been introduced into clinical practice, which can be used for ADLV common collector narrowing: the essence of the method consists in endovascular balloon dilatation of the narrowed section of the common collector. The general principles of radical correction of full abnormal pulmonary vein occlusion are reduced to the creation of a wide anastomosis of the pulmonary veins with the LP, closure of the ASD and ligation of the LV collector. The methods of surgical treatment vary depending on the anatomical form of total anomalous pulmonary vein occlusion. The first successful operation of connecting the pulmonary vein collector to the LV

was performed by W.Muller et al. Since the operation was performed before the "open heart" era, the ASD was not eliminated. Further, in 1955 F. Lewis reported about radical correction of malformation using subsurface hypothermia - there was anastomosis between common LV and LV collector with intracranial jamming of the latter, and ASD was eliminated via PP. Under hypothermic conditions the first successful radical surgery was performed by J.Kirklin et al. and D.Cooley and A.Oshner.

Radical correction is usually performed under IC conditions, and in newborns deep hypothermia with circulatory arrest is also used. There are two surgical options. The first variant is indicated for severe LV hypoplasia: to prevent postoperative cardiac insufficiency, ASF is liquidated partially using perforated patch. Two-stage approach is possible: the first stage is anastomosis between the common LV and LP; after the development of the left parts, the second stage is ligation of the common collector at the place where it enters the systemic vein. The second option is simultaneous correction of all components of the defect under IR conditions.

For correction of full abnormal pulmonary veins I and III according to R.Darling' et al., characterized by the presence of a common LV collector, similar operations are performed. The operation is usually performed from the median sternotomy, under IC conditions. The anastomosis between the LP and the LV collector can be performed by access to the heart through the PP and extracardiacally. The anastomosis area should not be less than the atrioventricular orifice, as small dimensions of the junction can lead to blood stasis in the LV; after anastomosis, the ASD is closed with a patch of autopericardium or synthetic tissue; and if the size of the BC cavity is small, the cavity can be enlarged by moving the lower edge of the patch toward the BC. Later, the operation was modified by N.M. Amosov et al. and B. Barrat-Boues et al., Back access to the LP suggested in 1970 by B. Roe was not recognized. Correction of cardiac type of malformation according to the method of J.Kirklin et al. In case the common pulmonary vein branch falls into BP, its orifice is usually wide, the correction consists only in relocation of the LV orifice into LP with simultaneous ASD plasty. When abnormal pulmonary vein inflow into the CS, a similar operation, modified by Van Praagh et al. is applied - first, a part of the septum between the oval window and CS is dissected, then the anterior wall is dissected, resulting in a newly created wide CS mouth being moved into the LP; the patch is sutured so that the CS mouth is moved into the LP cavity after its fixation. A frequent postoperative complication of this type of correction is conduction abnormality with various forms of bradyarrhythmia, which is apparently associated with intraatrial interstitial pathway damage during atrial septum resection. To prevent this complication, Van Praag et al suggested the

method, including CS "roof" resection with the following suturing of CS orifice and ASD in 1972. However, Reed G.E. et al. applied this surgical technique and noted no cases of bradycardia reduction in the postoperative period. At the same time, another serious complication of this modification was revealed: LV stenosis in the collector or orifice of CS, more often developing 3-4 months after the operation. Thus, the incidence of LV obstruction with complete anomalous pulmonary vein occlusion in CS after malformation correction, according to C.M.Whight et al., occurred in 3.6% of cases. Turley et al. observed pulmonary vein obstruction in 60% of cases, J.Davis et al. - in 10% and G.Reed et al. - in 22%.

Special technical difficulties usually arise in infracardiac and supracardiac correction with anomalous drainage not directly into one of the vena cava veins: a complicating issue in such cases is that LV and LP are far apart and therefore complex reconstructive surgeries are performed as recommended by several authors. In this case a tunnel carrying arterial blood is created in the VEP or LPV using a patch or longitudinal splitting of its cavity; this way the created canal intraatrial is moved through ASD into LV cavity.

In mixed type of malformation the method of corrective surgeries is more diverse. Depending on specific anatomical forms, it consists of a complex of operations performed for total and partial anomalous pulmonary vein occlusion. The success of all types of surgeries depends on creating an adequate anastomosis between the LP and pulmonary veins, careful prevention of air and arterial embolism.

Mortality after surgical correction of full abnormal pulmonary vein occlusion, according to some authors, ranged from 25 to 84%, and has recently decreased to 6%, and in young children it has been reduced to 16%. Significantly lower mortality can be achieved in children under 3 years of age. Lethality is usually associated with initial severe condition of the patients operated on against the background of severe LH and arterial hypoxemia. According to M. Gomes et al. the mortality rate after the surgery in children under 6 years old was 10%. , was 10%. It is higher in infracardiac form of abnormal pulmonary vein drainage in NPV, with LH, hypoplastic left heart.

LONG-TERM RESULTS. The study of long-term results is one of the most reliable indicators of the appropriateness of a particular treatment method, which allows assessing the technique from the point of view of time. Long-term results of surgical treatment of anomalous pulmonary vein occlusion are reported in the literature in single papers and more often in a small material.

Gomes et al. were among the first to pay close attention to the results of surgical correction in the long-term period after complete anomalous pulmonary vein occlusion. They studied 49 patients, out of 59 operated on, within 1 to 14 years. One

patient died 2 years after the operation from arrhythmia. Four patients underwent ACG study in the remote period with suspicion of residual shunt, and in two of them it was detected (6 and 14 years after surgery). The systolic pressure in the LA in all examined patients was within 30 mmHg.

Contradictory results after correction of full abnormal pulmonary vein occlusion were obtained by Trusler S.A. et al. Of 35 patients they operated on, twelve survived. Examination including cardiac cavity catheterization performed in them some years later showed good results. Only one patient with left lung aplasia had the LA pressure still equal to half of the systemic pressure. The authors found that children with subcardiac form of malformation had higher LA pressure than other types of Anomalogic Lung Venture, and this explained the reasons for unsatisfactory immediate results in their work.

According to A.Serra et al. and A.Serraf et al. , LV stenosis is a serious complication in patients before and after radical correction of full abnormal pulmonary vein occlusion. The authors present the results of surgery in 107 patients, 28 of them with subcardiac and 23 with supracardiac forms, out of which 57 patients had manifold stenosis. In the postoperative period 20 patients died (18%). Long-term follow-up of 4 patients revealed obstruction at the anastomosis site. One patient died without surgery out of two, reoperated, one also died in the postoperative period. In the last case the patient underwent balloon dilatation of the obstructed part. A similar case of successful transluminal balloon dilatation of the stenotic part of the colliculus in the patients after the operation was described by other authors as well.

A great experience in the treatment of complete abnormal pulmonary vein occlusion is described in R.Lamb et al. - 80 patients. In the long-term period there were 6 patients who died. The cause of five deaths was venous obstruction (5 weeks - 3 months after the operation). Stenosis of the CS orifice was seen in two cases (1 in mixed, the second - abnormal inflow of the pulmonary veins into the CS); stenosis of the LV manifold in two cases (supracardiac and subcardiac forms) and stenosis of the right LV orifice, resulting in right pulmonary edema, was in one case (cardiac form in PP). The LA pressure in these patients ranged from 65 to 120 mm Hg. The cause of death in the sixth patient was obstruction of the LPV with full abnormal pulmonary vein infiltration into the superior vena cava. Repeat surgeries were performed in 8 patients (three with obstruction, four with recanalization of the ASD and one with ASD obstruction). One of the conclusions made by the authors is that the technique of correction in full abnormal pulmonary vein occlusion in CS suggested by R.Van Praagh et al. is controversial.

Friesen C.L.H. et al. describe long-term results in 28 patients up to 14 years. Stenosis of the venous collector was detected in five patients, which required repeated surgical intervention (in three cases the operations ended in death). Recanalization of the ASD was another reason for repeated operations in 4 patients. As reported by Y.Ando et al., in the long-term period, out of 38 neonates operated with supracardiac complete abnormal pulmonary vein infiltration by Gerson technique, there were three cases of narrowing of the anastomosis site, two of which ended in the patient's death. A. corpo et al. achieved good long-term results up to 6 years in 30 patients. They used the so-called "double patch" technique - on the ASD and obligatorily a patch dilating the LP. There were no deaths in the postoperative period and all patients felt well.

To date, reports on errors during surgery (incomplete correction of the defect) and complications in the postoperative long-term period (septic endocarditis, leading to partial or complete recanalization of the defect) continue to appear in the literature. The possibility of defect recanalization is reported by many authors . Another group of specific complications in the long-term period is associated with obstruction of blood flow through the abnormally descending LV due to their narrowing, thrombosis. According to Friesen C.L.H. et al, 4 patients had signs of thrombosis of abnormally descending pulmonary veins with signs of lobe lung stasis, predominantly of the right upper lobe with hemoptysis in one patient. Such complication is seldom the cause of death but it doesn't pass away as the adhesions and collaterals develop, the blood from this lobe enters the systemic veins thus causing the arterio-venous outflow resumption.

There are specific complications associated with impaired blood flow along the VPV due to various reasons - narrowing of its lumen after separation, wrinkling of the patches, thrombosis. Husain S.A. et al. et al., in one patient noted signs of temporary occlusion of the VPH. C.Chartrand et al. found stenosis of the pulmonary veins 5 years after correction of abnormal pulmonary vein occlusion in one patient with augmented VPH. V.S. Sergievsky et al. recorded a case of thrombosis leading to cerebral edema. VEPH blood flow disorders can end safely and be compatible with life in spite of narrowing or even complete obstruction. Blood flow in this case is carried out by collaterals through the unpaired vein system in the NPV or through the accessory VEP, if there is one.

Based on the above, it is clear that there are many unresolved problems in the treatment of anomalous pulmonary vein drainage, among which are the danger of pulmonary-atrial anastomosis narrowing, ERV, pulmonary vein outflow disorder, left-right shunt preservation and heart rhythm disorders.

Conclusions

Thus, total anomalous pulmonary vein drainage is a rare and complex heart defect with rather typical clinical presentation and early development of pulmonary hypertension. ECHOCG, radiography, MSCT and cardiac catheterization with ACG play a leading role in the timely diagnosis of the malformation. Early and adequately performed reconstructive intervention with pulmonary vein dislocation into the left atrium provides good long-term results.

Literature:

1. Amosov N.M., Zynkovsky M.F., Ignatov P.I., Panichkin Y.V. Surgical treatment of total anomalous pulmonary vein drainage. Chest Surgery. -2019. -N1. -C.3-9//Амосов Н.М., Зынковский М.Ф, Игнатов П.И., Паничкин Ю.В. Хирургическое лечение полного аномального дренажа легочных вен. Грудная хирургия. -2019. -N1. -C.3-9.
2. Sergievsky V.S., Dugmanov E.K., Grenz V.G. Long-term results and rehabilitation of patients with secondary atrial septal defect. Cardiology. -2017. -N7. -C.32-37.//Сергиевский В.С., Дугманов Е.К., Гренц В.Г. Отдаленные результаты и реабилитация больных с вторичным дефектом межпредсердной перегородки. Кардиология. -2017. -N7. -C.32-37.//
3. Syrnev Y.A., Boboev A. K., Bondar V.Y. Surgical treatment of abnormal pulmonary vein drainage. Fifth All-Russian Congress of Cardiovascular Surgeons. Theses of reports and communications. Novosibirsk. November 23-26. -1999. p.20.//Сырнев Ю.А., Бобоев А. К., Бондарь В.Ю. Хирургическое лечение аномального дренажа легочных вен. Пятый Всероссийский съезд сердечно-сосудистых хирургов. Тезисы докладов и сообщений. Новосибирск. 23-26 ноября. -1999 г. - С.20.
4. . Obukhov V.N. Obukhov I. V. Once again about surgical treatment of abnormal pulmonary vein infiltration into the superior vena cava. IY-Всерс. Congress of Cardiovascular Surgery. 8-11 Dec. -1998. -C.30.
5. Обухов В.Н. Обухов И. В. Ещё раз о хирургическом лечении аномального впадения легочных вен в верхнюю полую вену. IY-Всеросс. съезд серд.-сосуд. хир. Тез. докл. и сообщ. Москва. 8-11 дек. -1998. -С.30
6. Ando H., Yasia H., Kado H. et al. Surgical repair of total anomalous pulmonare venous connection in neonates and infants-prevention for pulmonare venous distruction at the site of anastomosis. //Nippon Kyobu Geka Gakkai Zasshi -1990. -Vol. 38. -N.4. -P.518.

7. Ashoush R., Jebara V.A., el Rassi I., Badawi G., Sarcis A., As- mar B. et. al. Total anomalous pulmonary venous connection in adults; A surgical review. J.Med. Liban -1993. -V.41. -N.4. -P.230-235.
8. Bouharin V.A., Podzolkov V.P., Ilyin V.N. et. al. Correction of total anomalous pulmonary venous drainage in patients from in fancy to adolescence. The second World Congress of Ped. Cardiology I and Cardiac Surgery. Honolulu. Hawail. -1997. -P.372.
9. Bove E., de Leval M., Taylor J. et al. Infradiaphragmatic total anomalous pulmonary venous drainage: surgical treatment and long-term results. Ann. Thorac.Surg. -1981. -V.31. - N6. -P. 544-550.
10. Chartrand C., Payot M., Davidnon A. et. al. A new surgical approach for correction of partial anomalous pulmonary venous drainage into the SVC. J. Th. Cardiovasc. Surg.-1976. -V.71. -N.3. -P.29-34.
11. Corno A., Giamberti A., Carotti A. et. al. Total anomalous pulmonary venous connection surgical repair with a double-patch techique. Ann. Thorac. Surg. -1990. -V.49. -P.492-94.
12. Cooley D., Ochsner A. Correction of total annomalous pulmonary ven drainage tech. considerat. Surg. -1957. -V.42. –P.1014-21,
13. Darling R.S., Rothney M.B., Greid J.M. Total anomalous pulmonary venous return into the right side of the heart. Lab. Invest. -1957, -V.6. -N.1, -P.44-64.
14. Davis J., Ehrlich R., Hennessey T. Long-term follow-up of cardiac rhythm in repaired total anomalous pulmonary venous drainage. Thorac. Cardiovasc. Surg. -1986. -V.34. -N.3. -P.172.
15. Ehrenhaft J., Theilen E., Lawrence M. The surgical treatment of partial and total anomalous pulmonary venous connections. Ann. Surg. -1958. -V.148. -N.2. -P.249-258.
16. Friedli B., Guerin R., Davignnon A., Fouron J.C. Surgical treatment of partial anomalous pulmonary venous drainage: A long-term fallow-up study. Circulation.-1972. -V.55. -P.159-70.
17. Fukahara K., Murakami A., Ueda T. et.al. Usefulness of MRI and cine MRI in the assessment of TAPVC. The second World Congress of Ped. Cardiology and cardiac Surgery. Honolulu. Hawail. -1997. –P.344.
18. Gomes M., Feldt R., McGoon D et al. Long-term results following correction of total anomalous pulmonary venous connection. J. Thorac. Cardiovasc. Surg. -1971. -V.62. -N.2. -P.253.
19. Kirklin J.W., Ellis F.H., Wood E.H. Treatment of anomalous pulmonary venous connections in association with interatrial communications. Surgery. -1956. -V.39.

-P.389-98.
20. Lamb R.K., Qureshi Sh.H., Wilkinson J.L. et.al. Total anomalous pulmonary drainage seventeen year surgical experience. J.Thorac. Cardiovasc. Surg. -1988. -V.96. -N.3. -P.368-75.
21. Lewis F. The surgical anatomy of atrial septal defects: experiences with repair under direct vision. Ann.Surg. -1955. -V.142. -N.3. -P.401-414.
22. Miller W., Rashkind W., Miller R. et al. Total anomalous pulmonary venous return: effective palliation of critical ill infants by ballon atrial septostomy. Circulation. -1967. -V.35. -Suppl.2. -P.11-189.
23. Me Manus B.M., Leutreler J., Roberts W.C. Total anomalous pulmonary venous connection: Survival for 62 years without surgical intervention. Amer.Heart. J. -1982. -V. 103. -N.2. -P.298-301.
24. Reed G.E., Clauss R.H., Tice D.A. Correction of anomalous pulmonary venous drainage to the superior vena cava. Surg. Gynecal Obstet. -1962. -V.115. -P.771.
25. Serra A., Bruniaux J., Lacour-Gayet F. et al. Obstruction of total anomalous pulmonary venous drainage. Factors influencing improvement in surgical results. Arch.Mai.Coeur. -1990. -V.83. -N.5. -P.643-646.
26. Serraf A., Bruniaux J., Locour-Gayet F. et.al. Obstructed total anomalous pulmonary venous return. Toward neutralization of a major risk factor. J. Thorac. Cardiovasc. Surg. -1991. -V.101. -P.301-606.
27. Trusler C.A., Kazanelson G., Freedom R.N. Late results fallowing Repair of partial anomalous pulmonary venous connection with sinus venosus asd. J.Th. Cardiovasc. Surg. -1980. -V.79. -N.5. -P.776-781,
28. Turley K., Tickez W.Y., Ullyot D.J., Ebert P.A. Total anomalous pulmonary venous connection in infancy: influence of age and type of lessions. Amer.J.CardioL. -1980. -V.45. -P.92-97.
29. Van Praagh R., Harken A., Delisle G. et al. Total anomalous pulmonary venous drainage to the coronary sinus. J .Thorac. Cardioivasc. Surg. -1972. -V.54. -P.132
30. Van de Wal H.J.C.M. Total anomalous pulmonary venous drainage by double connection corrected ”y”’ ascending vein and coronary sinus repair. J.Th.Cardiovasc. Surg. -1993. -V.105. -N.2. -P.367-368.