CLINICAL CASE

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THE OUTCOME OF PRENATALLY DIAGNOSED CONGENITAL HEART DISEASE – AORTIC ARCH DOUBLING

ABSTRACT

The article is devoted to contemporary diagnostics of rare developmental anomaly of cardiovascular system – aortic arch doubling. The clinical case of early diagnostics of aortic arch doubling during the period of screening of pregnant woman in term of 22nd gestation week is considered. No actual frequency of the pathology is known, as 90 % of cases can be symptomatic. Among patients with CHD hospitalized in the cardiosurgical clinics a vascular ring is registered less than at 1 %. Approximately 50 % of these patients have trachea congenital anomaly such as absence of membranous area in tracheal cartilages. As a result, cartilages make a complete ring that aggravates tracheal stenosis. When esophagus considerably compressed in the first days of life it can be diagnosed as esophageal atresia. Swallowing problem is not constant, weakening in some cases, even disappearing and appearing again. Since birth the patient G. in a clinical picture had dominant trachea narrowing manifested with mixed stridor. Symptoms of dysphagia appeared on 4-5th months of life. No hemodynamic abnormalities at the patient were observed. The computer tomography data were proved to be relevant after the operative intervention. When analyzing the follow up study the newborn's arrested physical development was revealed, whereas psychomotor development was normal. Within the year there was obstruction of respiratory ways, also symptoms of dysphagia. The early diagnosis of the congenital heart disease subjected to correct tactics of the patient management since the birth, accelerating the process of surgical intervention as well as reducing the risk of life-threatening complications.

Keywords: a newborn, congenital heart disease, aortic arch doubling, dysphagia, breath

INTRODUCTION

Aortic arch doubling (AAD) is the rare anomaly at which aorta covers trachea and esophagus like a ring, it making up 1 % from number of all CHD [2]. Prenatally the diagnosis is revealed at 0,01 % of all surveyed. At the same time, frequency of operations concerning vascular ring among the population makes up 0,0013 %. Thus, for 90 % the pathology can proceed without symptoms [1].

As an isolated defect the aortic arch anomalies are noted in the formation of vascular ring along esophagus and trachea, causing the compression of the above-stated bodies. There are two types AAD such as functional aortic arch doubling and aortic arch doubling with left arch atresia, the latter noted as the rare pathology. At all these anomalies there are no hemodynamic abnormalities, this feature being the main distinction from other congenital heart diseases and great vessels [4, 5]. Cardiac tones are pure, noise is absent. The symptoms depend on compression of abnormally passing aorta and its branches to esophagus, trachea and recurrent nerve. They are manifested as difficulties in breathing and swallowing, phonation disturbance, sometimes they can be absent or present depending on a case. Dysphagia is one of characteristic signs. Approximately 50 % of these patients have the congenital anomaly of trachea such as absence of membranous area in tracheal cartilages. As the result cartilages make the complete ring that aggravates trachea stenosis [3, 5]. Considerable narrowing of trachea is revealed more often in the mixture with stridor. Concomitant severe tracheomalacia aggravates the prognosis. Difficulty in the diagnostics is caused not only by lower incidence rate, but also by lack of objective information on the pathology in periodical press [3].

MATERIALS AND METHODS

Clinical follow-up study of the patient with CHD, AAD in the neonatal pathology unit №1 (NPU №1) in 2016 in the Prenatal centre SAE RS (Y) «RHN№1-NCM».

RESULTS OF RESEARCH

The patient G. at the five-day age was hospitalized in NPU №1 with complaints on noisy strident breath at crying.

According to the anamnesis the child's mother is 31 years old, 5th pregnancy, toxicosis and ARVI at first trimester. The childbirth at term, 3 medical abortions, chlamydial and cytomegalovirus infections. On the second prenatal investigation (20 weeks of pregnancy) CHD AAD was diagnosed at the fetus. Trisomy 21, 18, 13 syndromes out of the amniotic fluid were excluded after the DNA testing. The second childbirth, at term, cephalic presentation. The newborn was a live boy, in a severe state with Apgar 7/9, full-sized, sanitation of upper airways in the maternity hall and further was taken to the intensive therapy department. Physical, morphofunctional maturity corresponded his gestation age (weight 3300g, height 51 cm, head circumference 34 cm, breast circumference 33 cm). In the neonatal pathology unit saturation decrease on 74 at rest was noted. When crying at birth inspiratory stridor was marked. In the first days of life moist rales were listened at auscultation . On phenotype the child had some abnormalities: relative disproportion due to slightly truncated limbs, ocular hypertelorism, wide philtrum, microgenia, short neck, four-finger fold on both palms. In the neurologic status a little reduction of muscular tone and depletion of inborn reflex are revealed. 1st stage of the investigation included electrocardiogram, ECHO cardiogram, X-ray of thoracic organs in direct projection, x-ray computer tomography of lungs with contrast.

The electrocardiogram was conducted on the second day of life, sinus rhythm with heart rate 143 beats per minute. Electric cardiac axis is rejected to the right. Signs of overstrain (hypertrophy) of the right ventricle. Infringement of repolarization processes.

The X-ray of thoracic organs at birth revealed pulmonary fields of mild transparency at the expense of hypoventilation, detected mostly in medial parts of both sides. Few structures in lung markings. Roots of lungs are not differentiated. Cardiac shadow is of typical size and form. Upper mediastinal widening is noted. Pulmonary-pleural sinuses are free. Diaphragm contours are even and accurate. Slight pneumaticity on the part of abdominal cavity organs. In dynamics, for the third days of life hypoventilation on the right was remaining.

The ECHO CG data did not exclude CHD and recommended RCT. It revealed interatrial septum aneurysm with shunting d=0,38. Functioning arterial channel (d=0,18 cm). Dilatation of the

right atrium (2,0 cm), right ventricle cavity (1,1 cm), pulmonary trunk (0,9 cm). Additional trabecular in the left ventricular cavity. EF - 71 %. Double aortic arch is not excluded.

According to the RCT of lungs aortic arch doubling is revealed. Aneurismal outpouching of distal part of the left aortic arch is marked, aortal diverticula is not excluded and the clinical diagnosis CHD is disclosed. Aortic arch doubling. Interatrial septum aneurysm with shunting. Functioning arterial channel. Impaired circulation of 1 stage. Respiratory insufficiency of 0-I stages.

The NPU №1 was supervising the case. Jaundice syndrome began on the third day, the maximum rate was 235 mkmol/l in 7 days. The child was on demand chest feeding at all stages, the weight value with positive dynamics (growth on 208 g for 1 week). In act of breastfeeding no difficulties in swallowing were noted. Stridor cry remained. The symptomatic treatment was carried out. In the hospital the boy was examined by the geneticist, cardiologist, oculist and the neurologist. After the correspondence consultation on neonatal cardiology (14.00.009) for performing highly technological aid the child was hospitalized in the Scientific institute of cardiovasculatory pathology named after E.N. Meshalkin, Novosibirsk.

When admitted in the scientific institute there were complaints to atony, dyspnea, hyperhidrosis. According to the multispiral computer tomography the diagnosis aortic arch doubling was proved, in addition, dominant left aortal arch, 2,2x3,8 mm trachea constriction, the diagnosis CHD. Aortic arch doubling with esophagus and trachea compression: dominant left aortic arch. Distal segment obstruction of the right aortic arch. Interatrial septum aneurysm. Patent foramen ovale.

On 18th day of life the patient was operated by conducting a section of aortic arch with separation of the vascular ring and trachea and esophagus decompression. Ligation with transecting patent ductus arteriosus. The postoperative period was critical due to respiratory insufficiency, ALV was used within 18 hours. Antibacterial therapy was carried out including Cephalosporin and Meropenem, diuretics, broncholytics, infusion-transfusion therapy, physiotherapy, massage. At 1 month and 11 days the child was discharged in a state of moderate severity, with preservation of stridor breath, slight dyspnea at rest, tube feeding as a result of breastfeeding refusal, frequent vomiting provoked by cough.

The follow-up data: at one year disharmonious development and hypotrophy of 2 stage was noted at the child's physical development. According to L.T.Zhurba, E.M.Mastjukova's psychomotor assessment (1981) the total score was 27 regarded as age norm. At one-year-old age stridor breath remained at rest, dysphagia was periodically observed. The child had twice pneumonia, ARVI. Preventive vaccinations were cancelled till 18 months.

CONCLUSION

Rare cardiovascular abnormalities require precise attention at their diagnostics. Even when CHD diagnosed, due to absence of characteristic clinical symptoms the diagnostics can be incomplete, thereafter resulting in serious consequences. Qualitative prenatal diagnostics promotes the earlier diagnosis of rare developmental anomalies in the neonatal period and leads more favorable prognosis for the disease. According to radiological and ultrasonic research all the patients with the syndrome of trachea and esophagus compression particularly the patients with signs of aortic arch anomaly were shown the RCT data for acknowledgement or exception of that diagnosis.

REFERENCES:

1. Arakelyan V.S. Ivanov A.A. Makarenko V.N. Problemy diagnostiki vrojdennoi deformatsii dugi aorty [Issues of diagnostics of aortal arch congenital abnormalities] Detskie bolezni serdtsa i sosudov [Children's cardiovascular diseases]. 2010, №3, pp. 31 - 37.

2. Arakelyan V.S. Gidaspov N.A. Kulichkov P.P. Dvojnaja duga aorty s formirovaniem sosudistogo kol'tsa i razvitiem sindroma kompressii trakhei i pischevoda: osobennosti klinicheskoi kartiny, diagnostiki i khirurgisheskoi taktiki [Aortal arch doubling with formation of vascular ring and development of a syndrome of trachea and esophagus compression: characteristics of clinical picture, diagnostics and surgical tactics] Grudnaya i serdechno-sosudistaya khirurgiya [Chest and cardiovascular surgery]. 2016, №5, pp. 299 - 302.

3. Bokeriya L.A. Arakelyan V.V. Vrojdennye anomalii dugi aorty. Diagnostika, taktika lecheniya [Congenital anomalies of aortal arch. Diagnostics, management] Grudnaya i serdechno-sosudistaya khirurgiya [Chest and cardiovascular surgery]. 2012, №4, pp. 14 - 19.

4. Gidaspov N.A. Taktika i neposredstvennye resul'taty khirurgicheskogo lecheniya patsientov s anomaliyami dugi aorty pri ikh sochetanii s drugimi zabolevaniyami grudnoi aorty i ee vetvei. Avtoref. kand … med. nauk [Management and short-term results of surgical treatment of patients with aortal arch anomalies at their combination to other diseases of chest aorta and its branches: thesis of cand… medical sciences] 14.00.44. Moscow, 2009, pp.24.

5. Krivchenya D.Y. Krivchenya A.K. Slepov T.D. Vnutrigrudinnye kompressii dykhatel'nykh putei, obuslovlennye dvoinoi dugoi aorty, u detei [Intrasternal compressions of respiratory ways caused by double aortal arch at children. Vrachebnoe delo [Medical practice]. 2006, №5, pp. 30 - 36.

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