Prenatal diagnosis of the fetal common arterial trunk. A case series

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Abstract

Fetal common arterial trunk is an anomaly represented by a unique arterial trunk that arises from the base of the heart, and gives birth to systemic branches, both pulmonary and coronary, frequently associated with a ventricular septal defect (VSD) and has a poor prognosis. We present a series of 17 cases diagnosed in our tertiary center with different types of fetal common arterial trunk, its associated disorders, the evolution of the pregnancies, and of the neonates. We concluded that our cases support the fact that a complete intrauterine evaluation of each case of the common arterial trunk is impossible. The postnatal prognosis of the cases from our center was fatal, similar to most reports of the literature.

Keywords: common arterial trunk (CAT); intrauterine diagnosis; evaluation; prognosis

Introduction

Fetal common arterial trunk (CAT) is a congenital anomaly represented by a unique arterial trunk that arises from the base of the heart, gives birth to systemic branches, both pulmonary and coronary, being frequently associated with a ventricular septal defect (VSD). This fetal anomaly carries a poor prognosis, and, according to Collett et al [1] CAT is divided into 4 types: type 1 – pulmonary trunk, with two branches that originate from the truncus arteriosus; type 2 – branches originating from the same level of truncus arteriosus (posteriorly); type 3 – the branches originate from different levels of the same truncus arteriosus (laterally); and type 4 – branches originate from the descending aorta or the aortic arc (newly called atresia of the pulmonary valve with VSD). The classification of Van Praagh et al [2] comprises: type A1 with partially separated pulmonary trunk; A2 with 2 branches that originate separately directly from the truncus arteriosus; A3 with a single pulmonary branch that originates directly from the truncus arteriosus and collateral vessels from the descending aorta; and A4 consist of anomalies of the aortic arc associated with truncus arteriosus. The classification of the Society of Thoracic Surgeons admits only 3 types of CAT from the classification of Van Praagh – aortic dominant, non-confluent type, and pulmonary dominant [3].

Prenatal diagnosis and follow-up of CAT is important due to the frequent association with other intracardiac abnormalities or complications such as truncal valve stenosis and insufficiency, which lead to cardiac failure, hydrops, and intrauterine death [4]. The survival of CAT in infancy is possible only with a correct surgical correction [1]. In addition, O’Byrne underlined that patients with CAT and reconstructive surgery presented important associated comorbidities, multiple re-interventions on catheter, a poor life quality, and deficit in exercise performance and in functional status [5]. The most frequent CAT is type 1 with a percentage of 60%, followed by type 2 with 35%, and type 3 and 4 with 5% [6].

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The aim of our study was to assess the correct diagnosis of the fetal CAT type, the gestational age at the time of the diagnosis, the anomaly’s associations and also the fetal and neonatal evolution in our series of cases.

**Material and method**

We performed a prospective descriptive study on 17 fetuses diagnosed in our tertiary center with CAT between February 2009 and April 2017. During this period, 621 pregnant women were referred to our center with the suspicion of fetal cardiac anomalies and 411 were confirmed, 17 of them having CAT. The 17 fetuses included in this group (4.13% from all structural cardiac anomalies) were diagnosed at a gestational age ranging between 15 and 36 weeks, among which only 12 of them had a gestational age lower than 24 weeks.

**Echocardiographic examination**

The fetal echocardiography was performed by a team with experience regarding fetal and neonatal cardiac anomalies, using Voluson 730 Pro (General Electric) or Voluson E8 Expert ultrasound machine with a RAB 4-8 MHz abdominal transducer. Data such as truncal valve regurgitation, type of pulmonary vessels emergency from a unique vessel and their prognostic implications were assessed. Ultrasound signs of other fetal anomalies were also evaluated within this group. Each pregnant woman was counseled by a pediatric cardiologist and a surgeon specialist in newborn cardiac surgery. The postnatal evaluation of the 4 newborns was made by a pediatric cardiologist in the first hours after delivery, using a Philips ultrasound machine iE 33 with a S8-3 transducer.

All the pregnant women signed written informed consent before inclusion in the study and we obtained the approval of the local Ethic Committee.

**Results**

We found CAT type 1 in 9 cases, type 2 in 2 cases, and in 6 cases we could not establish a well-defined type (fig 1). All the data about these cases are detailed in table I.

**Discussions**

The prenatal diagnosis of fetal CAT represents a challenge for obstetric physicians and pediatric cardiologists, and the difficulties in interpretation of the ultrasonography findings explain the increased misdiagnosing rate. Thus, the study of Swanson et al underlines a low rate of antenatal diagnostic of CAT, of only 32% out of the total number of patients diagnosed after birth with CAT [7]. The most common cardiac abnormalities misdiagnosed by both the fetal specialist imaging and pediatric cardiologist included pulmonary stenosis, VSD, myxoma, truncus arteriosus, and coarctation of the aorta [8].

Lee et al also emphasize that the confusion of CAT with aortic or pulmonary valve atresia is also commonly encountered [9]. According to these authors, out of the 17 fetuses that were prenatally diagnosed with CAT and underwent a postnatal echocardiography or autopsy, only 12 were confirmed to have CAT, 5 being incorrectly labeled as CAT, among which 3 presented pulmonary atresia and 2 associated aortic atresia [9].

Truncus arteriosus usually comprises a unique, dysplastic valve, with regurgitation which is unusual sten-
otic, with a number of 3 to 6 valves [6,10,11]. Almost 50% of the cases diagnosed with this anomaly do not present ductus arteriosus [12]. In our series of cases, regurgitation of truncal valve was encountered only in 2 cases, similar to the data reported by Volpe et al, where 4 cases out of 23 presented this association [13].

The differential diagnosis of CAT comprises the tetralogy of Fallot and pulmonary atresia with VSD, especially for types 2 and 3 [14]. In addition, the differentiation between the types of CAT is very difficult in the second trimester of pregnancy [13,15], and the accurate diagnosis within this gestational period represents a challenge even if MRI is performed [15]. Similarly, in our series of cases, we could not define the exact type of CAT in 41.17% of the cases (7 out 17), or name the exact origin of all arteries/pulmonary branches. It is also worth mentioning that for types 2 and 3 of Collett classification, the pulmonary branches might be impossible to be identified by ultrasound exam, even after birth requiring an angio-CT.

The most frequently encountered cardiac anomalies associated with CAT are aortic arch on the right side, ASD (atrio-septal defect), anomalies of the aortic arch branches, coarctation of the aorta, AVSD (atrio-ventricular septal defect), mitral atresia, and anomalies of the cardiac situs [6]. There were also reported several genetic disorders associated with CAT, such as: DiGeorge syndrome in approximatively 40% of the cases [16-21], trisomies 21, 18, or 13 in 4.5% of the cases [22,23], or even rare cases of trisomy 8 [24]. In addition, extracardiac malformations, such as genito-urinary abnormalities were also described in over 40% of the cases [19,21,22]. Similarly, Patel et al reported that out of 554 newborns diagnosed with truncus arteriosus, 204 cases presented the coexistence of noncardiac congenital anatomic abnormalities, genetic abnormalities, and syndromes (36.8%) [25]. In our study, associated disorders were found in 5 of the 17 cases (29.41%) diagnosed with CAT, and we also identified an association with trisomy 18 in one case.

The intrauterine prognosis of CAT depends on the insufficient or stenotic truncal valve that can lead to severe consequences such as: cardiac failure, hydrops or even fetal death [23]. Prenatal diagnosis of CAT remains challenging and is associated with a high rate of therapeutic elective termination [7]. Similarly, 8 out of the 17 pregnancies included in our study, were terminated through a therapeutic abortion, and probably those lost from our evidence reached the same course or died after birth.

The postnatal prognosis depends on multiple factors, such as: the anomaly type, the origin of the pulmonary branches, etc. According to a review which assessed the post-surgical evolution of CAT performed within the first months of life, only 28 out of 48 alive newborns with CAT survived [4], underlining that the diagnosis of CAT carries only approximately 50% survival chances. Another study pointed out a survival rate much lower.

<table>
<thead>
<tr>
<th>Case</th>
<th>GA</th>
<th>Screening diagnosis</th>
<th>CAT type</th>
<th>Amniocentesis</th>
<th>Ultrasound associations</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>21</td>
<td>CAT</td>
<td>1</td>
<td>declined</td>
<td>No</td>
<td>Newborn’s surgery/ died at 8 month, no genetic analysis</td>
</tr>
<tr>
<td>2</td>
<td>36</td>
<td>Cardiac anomalies</td>
<td>1</td>
<td>-</td>
<td>No</td>
<td>Lost of evidence</td>
</tr>
<tr>
<td>3</td>
<td>28</td>
<td>Cardiac anomalies</td>
<td>2/3*</td>
<td>declined</td>
<td>Tymus hipoplasia</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>4</td>
<td>22</td>
<td>CAT</td>
<td>2/3*</td>
<td>declined</td>
<td>Bilateral pielectasy</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>5</td>
<td>23</td>
<td>CAT</td>
<td>2/3*</td>
<td>declined</td>
<td>No</td>
<td>Lost of evidence</td>
</tr>
<tr>
<td>6</td>
<td>35</td>
<td>Cardiac anomalies</td>
<td>2/3*</td>
<td>declined</td>
<td>Retroghnatism</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>7</td>
<td>21</td>
<td>Cardiac anomalies</td>
<td>2/3*</td>
<td>declined</td>
<td>No</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>8</td>
<td>19</td>
<td>Cardiac anomalies</td>
<td>2/3*</td>
<td>declined</td>
<td>Retroghnatism, short femur</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>9</td>
<td>15</td>
<td>CAT Type 1</td>
<td>1</td>
<td>declined</td>
<td>Truncal valve regurgitation</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>10</td>
<td>22</td>
<td>CAT</td>
<td>1</td>
<td>normal karyotype</td>
<td>No</td>
<td>Newborn’s surgery/ died at 4 month</td>
</tr>
<tr>
<td>11</td>
<td>28</td>
<td>Cardiac anomaly</td>
<td>1</td>
<td>normal karyotype</td>
<td>Truncal valve regurgitation</td>
<td>Lost of evidence</td>
</tr>
<tr>
<td>12</td>
<td>22</td>
<td>CAT</td>
<td>1</td>
<td>normal karyotype</td>
<td>No</td>
<td>Alive newborn at 2 months after first step of surgery</td>
</tr>
<tr>
<td>13</td>
<td>23</td>
<td>CAT</td>
<td>1</td>
<td>declined</td>
<td>No</td>
<td>Therapeutic abortion</td>
</tr>
<tr>
<td>14</td>
<td>22</td>
<td>CAT</td>
<td>1</td>
<td>declined</td>
<td>Truncal valve regurgitation</td>
<td>Ongoing bichorionic pregnancy, one fetus with normal hearth</td>
</tr>
<tr>
<td>15</td>
<td>19</td>
<td>CAT</td>
<td>2</td>
<td>normal karyotype</td>
<td>No</td>
<td>Therapeutic abortion</td>
</tr>
</tbody>
</table>

GA – gestational age (weeks); CAT – Common arterial trunk; *type 2 or 3 could not be correctly diagnosed in utero
only 10% after surgical procedure for CAT [7]. The same study stated that even though the prenatal diagnosis led to a surgical repair at a younger age, the neonatal survival rate remained constant [7]. Nevertheless, though the survival rate after surgery is relatively low, it improves further in life. Therefore, Jacobs et al. underlined that the overall aggregate operative mortality after CAT repair was 9.4% [26], while Naimo et al. emphasized that the survival rate at 30 years reached 73.6% [27]. The patients with the most severe complex defects usually have the highest risk for death [28], raising also medical ethics issues [29]. Unfortunately, two of the three newborns from our center was fatal, similar to most reports in the literature.

The main limitation of our study is represented by the small number of cases. In these conditions in our study, we had an unfavorable prognosis of this anomaly. Therefore, we can conclude that, without taking into account the ongoing pregnancy with CAT, in our evidence there is just one alive newborn out of the 16 fetuses diagnosed in utero.

Conclusions

Our series of cases supports the fact that a complete intrauterine evaluation of each case of common arterial trunk is impossible. The postnatal prognosis of the cases from our center was fatal, similar to most reports in the literature.

Conflict of interest: None.

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References