The Human Fetal Venous System

Normal Embryologic, Anatomic, and Physiologic Characteristics and Developmental Abnormalities

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Objective. The introduction of high-resolution ultrasonography combined with color-coded Doppler imaging offered a breakthrough in the evaluation of the human fetal venous system, considerably enhancing our understanding of fetal venous circulation in normal physiologic conditions, as well as providing us the ability to study circulatory changes in abnormal circumstances. The purpose of this study was to describe the normal anatomic development and complex of anomalies of the human fetal venous system and to review recently published series of these anomalies. Methods. Normal embryologic and anatomic development is described. An English language literature search of recent MEDLINE listings was performed to glean data from recently published series reporting prenatal diagnosis of the various anomalies and their associated malformations. Results. Anomalies of the human fetal venous system occur sporadically, often associated with cardiac or other malformations. The pathophysiologic mechanisms leading to abnormal in utero development of the human venous system remain largely underdetermined. On the basis of the type of vein involved, embryologic precursor, and etiologic correlation (primary or secondary), classification into 4 major groups is described. Conclusions. Prenatal evaluation of fetuses found to have anomalies of the venous system should include a careful search for cardiac anomalies, including pulmonary venous drainage, and a detailed anatomic survey of the umbilical, portal, hepatic, and ductal systems to determine aberrant communication and, if possible, to discover clues to systemic diseases or thromboembolic phenomena. Key words: anomalies; fetal venous system; prenatal diagnosis.

Abbreviations
PAPVC, partial anomalous pulmonary venous connection; TAPVC, total anomalous pulmonary venous connection

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Recent developments in ultrasonographic imaging, including the development of high-resolution ultrasonography combined with color-coded Doppler imaging, facilitate prenatal diagnosis of fetal malformations of varying severity. This in turn enables caregivers to provide a prognosis to the parents depending on the malformation detected. Furthermore, prenatal diagnosis of fetal malformations, especially of the heart, has been found to improve morbidity and mortality rates during postnatal treatment.1–3

Although the fetal venous system was described almost 20 years ago,4,5 it was the introduction of high-resolution ultrasonography combined with color-coded Doppler imaging that offered a breakthrough in the evaluation of this system,6–8 considerably enhanc-
ing our understanding of fetal venous circulation in normal physiologic conditions as well as providing us the ability to study circulatory changes in abnormal circumstances. Nevertheless, the pathophysiologic mechanisms leading to in utero abnormal development remain largely undetermined.6

The most commonly reported abnormalities involve the intrahepatic umbilical vein, but rare cases of agenesis of the ductus venosus have also been reported.9–10 Improvement in imaging technology led to a search for the in utero diagnosis of other rare anomalies of the fetal venous inlet system.13–20 Two recent relatively large series describing fetal venous system malformations suggested that in a targeted fetal organ scan, the course of the umbilical vein, ductus venosus, portal and hepatic veins, inferior vena cava, and pulmonary veins, as well as associated anomalies, can be clearly visualized and accurately diagnosed with the use of two-dimensional gray scale and color Doppler imaging.16,18

The only information currently available regarding the etiology, importance, and prognosis of these abnormalities is derived from neonates19–21; however, fetal outcome cannot be extrapolated from data obtained from neonates with similar abnormalities. As the number of case reports regarding various fetal vein abnormalities increases,14,22–24 prenatal treatment of these patients may become problematic, and understanding the embryologic basis and pathophysiologic mechanisms of fetal venous inlet system abnormalities becomes increasingly important for accurate prenatal counseling.

In this review we provide a detailed description of the embryologic, anatomic, and physiologic characteristics of the normal human fetal venous system. In addition, we describe a classification system of associated developmental abnormalities of this system based on the type of vein involved.

Normal Development of the Human Fetal Venous System

Embryologic Characteristics

A system of 3 paired veins is found in the 4-week embryo, which includes the umbilical veins from the chorion, the vitelline veins from the yolk sac, and the cardinal veins from the body of the embryo itself, all of which open to the right and left horns of the sinus venosus of the heart. Any further development of the fetal venous system represents changes occurring in these symmetric venous systems (Fig. 1A).

The fetal liver and its development in the septum transversus play an important role in modifying the primitive vitelline and umbilical systems into their final shape. First the developing hepatic sinusoids link to both vitelline veins and then tap the umbilical vein at day 32. Each vitelline vein is then interrupted by the sinusoidal labyrinth to a distal segment extending from the yolk sac to the liver, which is finally converted into the portal vein and to a proximal segment extending from the liver to the heart, in which the right proximal stem represents the hepatic vein, whereas the left proximal and right distal vitelline veins atrophy and disappear (Fig. 1B).

The umbilical veins also undergo several changes. In the 5-mm embryo, the left umbilical vein becomes the dominant conduit of blood from the placenta, emptying into the left horn of the sinus venosus. Laterally expanding primitive left and right lobes of the liver contact the paired umbilical veins coursing close by, thus forming an anastomotic system among liver sinusoids, vitelline veins, and umbilical veins, causing the loss of connection between both umbilical veins with the fetal heart. Eventually, in the 6-mm embryo, the entire right umbilical vein and proximal segment of the left umbilical vein atrophy and soon disappear. Thus all placental blood enters the right atrium through the left distal umbilical vein, ductus venosus, and proximal right vitelline vein, which bypass the liver sinusoids. As development continues, in the 9-mm embryo, the left and right portal veins are already part of the left umbilical vein, whereas the hepatic vein and ductus venosus drain into the infra-cardiac portion of the inferior vena cava (Fig. 1C).

The cardinal veins form the main venous drainage system of the embryo body, with the anterior and posterior cardinal veins draining the embryonic cranial and caudal parts of the body, respectively, which then empty into a common cardinal vein, which is the third venous system entering the sinus venosus of the primitive heart. The left brachiocephalic vein is formed during the eighth week from the right anterior and right common cardinal veins, through left-to-right anastomosis, whereas the left anterior cardinal vein disappears. After the atrophy of the posterior cardinal veins, the subcardinal and supracardinal veins are formed. The latter, after division
in the renal area, form the azygos and hemi-azygos veins above this level, whereas below this level the left supracardinal vein degenerates, and the right supracardinal vein becomes the caudal part of the inferior vena cava. The upper segments of the inferior vena cava are derived from the subsupracardinal anastomosis at the renal area, the prerenal segment from the right subcardinal vein, and the hepatic segment from the proximal vitelline vein and the liver sinusoids.

The common pulmonary vein can be identified in a 4-mm embryo as an invagination of the dorsal wall of the left atrium. As the atrial cavity develops, the stem of the pulmonary vein is gradually incorporated into the left atrial wall, until finally the 2 right and left branches of the pulmonary stem enter the atrial cavity. In the human embryonic venous system, the left umbilical vein eventually remains the only vein from the placenta to the fetal heart, and the development of its connection and the ductus venosus (“the critical anastomosis”) is of primary importance in the development of the venous drainage system and supply of oxygenated blood to the fetus. The ductus venosus is unique to fetal life, arising from the vitelline vein within the liver (hepatic sinusoids). From this vessel arise the portal vein, the superior mesenteric vein, and the intrathoracic inferior vena cava, which joins the ductus venosus to the heart (the hepatocardiac portion; Fig. 1C).

**Anatomic Characteristics**

By the end of the first trimester, the umbilical vein transfers blood from the placenta to the fetus. Its intrafetal portion ascends by way of the falciform ligament to join the umbilical segment of the left portal vein. The umbilical vein and the left portal vein maintain the same diameter, larger than the right portal vein, which receives blood primarily recycled through the main portal vein. There is therefore a marked preference in both the quality and quantity of blood flowing through the left lobe of the liver, which is noticeably larger than the right. This situation is reversed in the adult after atrophy of the umbilical vein and the ductus venosus.

The left portal vein joins the anterior and posterior branches of the right portal vein through an almost 90° right turn. From this same “twist,” known as the “pars transversa,” arises the ductus venosus, which joins distally with the left hepat-
ic vein and the inferior vena cava just proximal to the entrance into the right atrium (Fig. 2). The existence of a “sphincter” that regulates blood flow through the ductus venosus to the heart remains a subject of debate, as does the question of oxygen concentration–dependent anatomic nervous control of the activity of this sphincter. Table 1 summarizes the correlation of embryonic structures in the venous and arterial systems with the equivalent adult anatomy.

**Physiologic Characteristics**

Principal vessels such as the ductus venosus, hepatic veins, and inferior vena cava facilitate the transfer of blood with the highest possible oxygen concentration to the fetal heart. However, the pathophysiologic mechanisms controlling the function of the human fetal venous circulatory system are only partially elucidated. Low placental resistance and improved cardiac contraction aid in the creation of a pressure gradient between the atria and ventricles that reduces the preload in the venous circulatory system and allows blood to flow toward the heart. This intimate link between the function of the venous vascular system and the other elements of the fetal circulation is shown physiologically during fetal breathing movements, which have a direct effect on venous return to the heart. Examination of the effect of fetal breathing movements on the circulation in the venous system revealed that changes in the pressure gradient between the intraabdominal and intrathoracic cavities during breathing movements caused subsequent changes in blood flow in the venous system. Specifically, during inspiration the pressure gradient rose from approximately 0 to 3 to approximately 22 mm Hg, causing an increase in the pressure gradient between the umbilical vein and the thoracic part of the ductus venosus and leading to a rise in flow velocity in the umbilical vein, whereas during expiration the opposite events occur. Other studies showed that changes in intrathoracic pressure caused by fetal breathing movements affect both venous return to the heart and the arterial system.

Changes in venous return to the fetal heart can also influence emptying of the placental bed, as well as systolic and diastolic blood flow in the arteries. A decrease in venous return causes a drop in placental bed drainage and filling of the heart ventricles, resulting in a drop in the end-diastolic volume of the arteries on the one hand and corresponding decline (through the Frank-Starling mechanism) in stroke volume of the heart on the other side of the system. As a result, these changes may cause an additional negative effect on venous return and flow in the umbilical vein.

**Developmental Abnormalities of the Human Fetal Venous System**

We speculate that the normal development of the fetal venous system may be disturbed by either of 2 different events: (1) primary failure to transform or to form the critical anastomosis and (2) secondary occlusion of an already transformed system. Thus, on the basis of the type of vein involved, embryologic precursor, and etiologic correlation (primary or secondary), we have classified them into 4 major groups. Table 2 summarizes this classification system.
Abnormal Connection of the Cardinal Veins

Heterotaxy syndromes represent a pure example of abnormal primary development of the venous system, with a mouse model existing that explains the transmission of situs abnormalities in polysplenia-asplenia syndromes. 31,32 Most of these syndromes are associated with complex anomalies, especially cardiac malformations that influence the prognosis appreciably; however, cases without notable cardiac involvement in which fetuses may survive have been reported. 33 It is in these cases that accurate prenatal diagnosis is essential to provide appropriate counseling to the parents. Apart from the complex heterotaxy syndromes, isolated anomalies of the cardinal veins have also been reported.

Absence of the inferior vena cava with azygos continuation results from primary failure of the right subcardinal vein to connect with the hepatic segment of the inferior vena cava. Instead, it shunts its blood directly into the right supracardinal vein. Hence, the bloodstream that forms the caudal part of the body reaches the heart by way of the azygos vein and superior vena cava. The persistent left superior vena cava is another example of primary failure to create anastomosis and to form the left brachiocephalic vein.

Abnormalities of the Umbilical Veins

Abnormalities of the portal and umbilical veins form the major group of fetal venous anomalies. These mainly result from primary failure to form the critical anastomosis, with an aberrant vessel shunted between the placenta and the systemic veins. 13,21,22,34 Several of these cases with an abnormal umbilical vein connection were reported to be associated with Noonan syndrome. Hydrops fetalis was a common finding in these fetuses, although the severity of the edema was out of proportion to the vascular anomaly. This raised the possibility that the finding was largely attributable to some other cause, perhaps a congenital generalized lymphangiectasia as manifested in Noonan syndrome.

White et al 35 offered probably the most convincing explanation of an embryologic basis for this rare anomaly, introducing the term “critical anastomosis.” According to this theory, through unidentified causes of embryologic maldevelopment, anastomosis between the umbilical and vitelline veins fails, and degeneration of both umbilical veins occurs in early embryonic life. As a result, venous blood flow from the placenta is blocked, which leads to the opening of vascular channels through anastomosis to the supracardinal components of the inferior vena cava. Another study showed that in a 7-mm pig embryo there was a connection between the primitive pelvic venous plexus that drains the lower limb bud and the embryonic umbilical vein. 36

Summarizing the results of these studies, it may be concluded that in a case of primary failure to form these critical anastomoses, the placenta venous return is rerouted to the systemic veins. Primary failure to form the critical anastomosis may involve the iliac vein, the superior...
vena cava, or a direct communication with the right atrium or the left horn of the sinus venosus (coronary sinus; Fig. 3). Noteworthy is a common feature of all the aforementioned cases: agenesis of the ductus venosus, a fundamental embryologic maldevelopment. Diagnosis of this entity is feasible today with the widespread use of prenatal ultrasonography. Although agenesis of the ductus venosus was first described more than 100 years ago, several cases of neonates with this anomaly have since been reported.37,38

According to the findings, 2 subgroups may be distinguished. The first includes fetuses in which the umbilical flow entirely bypasses the liver, connecting to the systemic venous circulation as described above, and the second includes fetuses in whom the umbilical vein drains into the portal vein, thus causing all the umbilical blood to pass through the hepatic sinusoids. In the former group, many reported cases had an abnormal connection of the umbilical vein to the iliac vein, or to the inferior vena cava in its infra-cardiac segment, and some were associated with pleural effusion, hydrops, and Noonan syndrome.21,22,34 In the latter, an adequate connection between the umbilical vein and the vitelline venous system was established. The umbilical vein drains properly into the portal vein but fails to establish a communication with the persistent proximal part of the right vitelline vein. In such cases, prolonged hyperperfusion of fetal liver sinusoids may induce prenatal portal hypertension.22,39

Although complete primary failure to form the critical anastomoses in both the left and right umbilical veins is very rare, partial failure to form the critical anastomosis may be more common. In these cases, in which partial Anastomosis between the hepatic sinusoids and umbilical vein fails to develop, blood is then diverted through other channels. Two main paths have been described, the most common of which is the persistent right umbilical vein anomaly (Fig. 4).10,16,40 This anomaly is characterized by failure of the right umbilical vein to form anastomosis with the right vitelline vein and degeneration of the left umbilical vein,9,41,42 whereas the second, much less common path involves the direct entry of the umbilical vein into the right atrium.15,16,43,44

Several pathophysiologic explanations for these aberrations have been proposed. Jeanty9 suggested that streaming of early flow through the right umbilical vein, primary or secondary to occlusion by thromboembolic events arising from the placenta, might cause this anomaly. In addition, in experiments involving rats, teratogenic agents such as retinoic acid and folate deficiency were found to induce primary failure to form anastomosis and consequently the right umbilical vein to remain patent.45 Echogenic foci found within the fetal liver suggest a thromboembolism occluding the ductus venosus or other veins, possibly a secondary formation of such an anomaly. In such cases of portal-ductal occlusion, oxygenated blood coming from the placenta is diverted into the already closed proximal left umbilical vein, causing reopening of this part of the umbilical vein and resulting in a persistent proximal left umbilical vein.46

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**Table 2. Proposed Classification System for Abnormal Connections of the Fetal Venous System With the Heart**

A. Cardinal vein
   a. Complex malformations, heterotaxic syndrome
   b. Isolated malformation

B. Umbilical veins
   a. Primary failure to create critical anastomoses
      i. Complete: abnormal connection of umbilical vein (venous shunt) into iliac vein, inferior vena cava, superior vena cava, and right atrium
      ii. Partial: persistent right umbilical vein with or without ductus venosus
   b. Secondary occlusion

C. Vitelline veins
   a. Primary failure to create critical anastomoses
      i. Complete agenesis of portal system
      ii. Partial agenesis of right or left portal branch (portosystemic shunt)

D. Anomalous pulmonary venous connection
   a. Total
   b. Partial

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**Anomalies of the Vitelline Veins**

Abnormalities of the vitelline veins are extremely rare, and only a few have been reported during fetal life.\(^{13,16}\) Primary failure to form the critical anastomosis may lead to complete agenesis of the portal system or partial agenesis of the right or left portal branch.

Complete absence of the portal system presents an extreme example of total failure of the vitelline veins to transform into the portal system, stemming from a primary failure to form critical anastomosis with hepatic sinusoids or umbilical veins. As a result, the enterohepatic circulation is disturbed, and the portal venous blood is shunted systemically. Mesenteric and splenic venous blood may drain into the renal veins, hepatic veins, or directly into the inferior vena cava.\(^{47-49}\) To our knowledge, ultrasonographic prenatal diagnosis of congenital absence of the portal system is rare and has only been described in a single case in which the in utero ultrasonographic appearance of total agenesis of the portal system included an intrahepatic aberrant vessel, absence of the portal system, and marked dilatation of the inferior vena cava.\(^{50}\)

Incomplete absence of the portal system or partial failure to form critical anastomosis may represent a more benign form of vitelline vein abnormalities. Partial primary failure to form critical anastomosis may result in agenesis of the right portal system, with a persistent left vitelline vein connected directly to the hepatic vein (portohepatic shunt) and an absence of the ductus venosus, with neonatal spontaneous resolution.

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**Figure 3.** Images from a patient referred at 22 gestational weeks for investigation of cardiomegaly. Fetal echocardiography revealed cardiomegaly with dextrocardia and anomalous venous return to the coronary sinus with agenesis of the ductus venosus. The diagnosis was confirmed postnatally. **A**, The solid arrow indicates an aberrant vessel; and dotted arrow, coronary sinus. **B**, The arrow indicates the right portal vein; and asterisk, stomach. **C**, The arrow indicates the dilated umbilical vein.

**Figure 4.** Persistent right umbilical vein anomaly. AO indicates aorta; GB, gallbladder; IVC, inferior vena cava; and UV, umbilical vein.
occurring later in some of these cases.\textsuperscript{16,51} In addition, to our knowledge, only 1 case with agenesis of the right and left portal veins associated with a portosystemic shunt has been reported prenatally.\textsuperscript{17}

**Anomalous Pulmonary Venous Connection**

Anomalous pulmonary venous connection may be separated into 2 subgroups: partial and total.

Partial anomalous pulmonary venous connection (PAPVC) involves 1 or more, but not all, of the pulmonary veins connecting to the right atrium or a tributary (Fig. 5). Usually these malformations are also accompanied by an atrial septal defect. Several variant manifestations of this anomaly have been reported and include abnormal insertions of the right pulmonary veins into the superior or inferior vena cava and the right atrium and entry of the left pulmonary veins into the left innominate vein.

**Figure 5.** Common forms of PAPVC. **A,** Anomalous connection of the right pulmonary veins (R.P.V.) to the superior vena cava (S.V.C.). A high or sinus venous defect is usual in this anomaly. **B,** Anomalous connections of the right pulmonary veins to the inferior vena cava (I.V.C.). The right lung commonly drains by 1 pulmonary vein without its usual anatomic divisions. Parenchymal abnormalities of the right lung are common, and the atrial septum is usually intact. **C,** Anomalous connection of the left pulmonary veins (L.P.V.) to the left innominate vein (L.Inn.V.) by way of a vertical vein (V.V.). An additional left-to-right shunt may occur through the atrial septal defect. **D,** Anomalous connection of the left pulmonary veins to the coronary sinus (C.S.). L.A. indicates left atrium; L.V., left ventricle; R.A., right atrium; and R.V. right ventricle. Reprinted by permission from *Heart Disease in Infants, Children, and Adolescents.*\textsuperscript{1}
Visualization of pulmonary veins connecting to the right-sided cardiac structure involved, which may be the superior vena cava, left innominate vein, or coronary sinus, is the basis of echocardiographic diagnosis of PAPVC. Also, a finding of right ventricular volume overload with an intact atrial septum may mean that a PAPVC anomaly is present. Doppler flow studies and color mapping are indispensable in identification and classification of this group of anomalies.

In total anomalous pulmonary venous connection (TAPVC) there is no connection between the pulmonary veins and the left atrium; thus the pulmonary veins drain into the right atrium or the systemic veins (Fig. 6). About one third of patients have an associated major anomaly such as cor biloculare, single ventricle, truncus arteriosus, transposition of the great arteries, pulmonary atresia, coarctation, hypoplastic left ventricle, and anomalies of the systemic veins, whereas the remaining two thirds have isolated TAPVC. Although TAPVC is a rare lesion in its isolated form, occurring in only about 1 per 17,000 live births, it is one of the few emergencies left in pediatric cardiologic practice. This is because neonates with obstructed pulmonary venous return can have severe decompensation soon after birth, and unlike other forms of congenital heart disease, maintaining ductal patency by using prostaglandins is of little help. On the other hand, the subset of patients with an anomalous connection to the coronary sinus does not tend to have urgent symptoms, because the drainage to this site is rarely obstructed.

Several classification systems for TAPVC have been proposed, depending on the level of the anomalous connection. In one system, TAPVC is classified into 4 types, whether the anomalous connection is at the supracardiac level (I), cardiac level (II), infracardiac level (III), or 2 or more of the above levels (IV). An alternate classification system divides this anomaly into 2 groups: supradiaphragmatic without pulmonary venous obstruction and infradiaphragmatic with pulmonary venous obstruction. An atrial septal defect or patent foramen ovale is generally present. The presence of an obstructive lesion in the pulmonary venous channel influences the hemodynamic state and clinical manifestation of TAPVC. This obstruction may occur at the interatrial septum or may be intrinsic or extrinsic to the anomalous venous channel.

The aim of echocardiographic evaluation of suspected TAPVC lies in locating the site of the connection of the common pulmonary vein, evaluating any obstruction of the pulmonary veins or vertical vein, and confirming or excluding associated cardiac lesions. All forms of TAPVC will have right ventricle volume overload, an enlarged right atrium, and a bowing leftward of the interatrial septum. Total anomalous pulmonary venous connection should be suspected when no pulmonary veins can be visualized entering the left atrium on the short-axis scan. In addition, the common pulmonary channel may be identified posterior to the left atrium, with the pulmonary veins draining into it. The channel may be followed to the site of its connection. In supracardiac forms of TAPVC, the channel will usually drain into an ascending vertical vein, which in turn may be followed to a dilated systemic venous structure, most commonly the left innominate vein or superior vena cava. Doppler studies reveal flow in this vertical vessel to be cephalad, as opposed to the caudal flow found in the superior vena cava.

In infradiaphragmatic forms of TAPVC, the connection is usually at the portal venous system but may also be to the hepatic veins. The pulmonary veins converge into a common channel, small and inferior to the left atrium above the diaphragm, or it may appear as a discrete chamber. If untreated, affected neonates will survive only a few days to approximately 4 months.

Doppler examination is invaluable in the diagnosis of all forms of TAPVC. The abnormal color flow signal may be followed to identify the anomalous connection as well as the direction and mean velocity of flow. Turbulence from any obstruction will produce a mosaic color jet that reveals its location, whereas pulsed or continuous wave Doppler imaging allows for quantification of the degree of obstruction.

The prognosis in cases of PAPVC and TAPVC depends on the presence and size of the interatrial connection, the presence of any obstructive lesion in the anomalous venous pathways, the overall state of the pulmonary vascular bed, and the presence of any cardiac or extracardiac malformations. In cases of TAPVC with obstruction in the anomalous venous channel, the neonate usually dies in the first weeks of life.

The most common anomalies of the fetal systemic venous system diagnosed prenatally that have been described recently are summarized in Table 3.
Figure 6. Common forms of TAPVC. A, TAPVC to the left innominate vein (L.Inn.V.) by way of a vertical vein (V.V.). B, TAPVC to the coronary sinus (C.S.). The pulmonary veins join to form a confluence designated the common pulmonary vein (C.P.V.), which connects to the coronary sinus. C, TAPVC to the right atrium. The left and right pulmonary veins (L.P.V. and R.P.V.) usually enter the right atrium separately. D, TAPVC to the portal vein (P.V.). The pulmonary veins form a confluence from which an anomalous channel arises. This connects to the portal vein, which communicates with the inferior vena cava (I.V.C.) by way of the ductus venosus (D.V.) or the hepatic sinusoids. L.A. indicates left atrium; L.H., left hepatic vein; L.P., left portal vein; L.V., left ventricle; R.A., right atrium; R.H., right hepatic vein; R.P., right portal vein; R.V., right ventricle; S.M.V., superior mesenteric vein; S.V., splenic vein; and S.V.C., superior vena cava. Reprinted by permission from Heart Disease in Infants, Children, and Adolescents.\textsuperscript{1}
### Table 3. Common Anomalies of the Venous System

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No. of Cases</th>
<th>Sonographic Findings/Associated Abnormalities</th>
<th>Outcome</th>
</tr>
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<tbody>
<tr>
<td><strong>Cardinal veins</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right and left isomerism</td>
<td>13</td>
<td>7, abnormalities of viscerocardiac status</td>
<td>3, TOP</td>
</tr>
<tr>
<td>Atkinson and Drant⁵⁴</td>
<td>8, RI</td>
<td>7, complete atrioventricular septal defect</td>
<td>10, delivered with all associated abnormalities</td>
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<tr>
<td></td>
<td>5, LI</td>
<td>5, interrupted inferior vena cava;</td>
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<td></td>
<td></td>
<td>4, dysrhythmias</td>
<td></td>
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<tr>
<td><strong>Heterotaxy syndromes</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Marton et al⁵⁵</td>
<td>13</td>
<td>Congenital heart disease</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>9, SA</td>
<td>10, AV canal</td>
<td></td>
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<tr>
<td></td>
<td>4, SIT</td>
<td>10, great vessel anomaly</td>
<td></td>
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<tr>
<td>Cesko et al⁵⁶</td>
<td>2</td>
<td>None</td>
<td>NA</td>
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<tr>
<td></td>
<td>1, SI</td>
<td>Transposition of the great vessels</td>
<td></td>
</tr>
<tr>
<td>Oztunc et al⁵⁷</td>
<td>1, SIT</td>
<td>Transposition of the great vessels</td>
<td>NA</td>
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<td><strong>Umbilical veins</strong></td>
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<td></td>
<td></td>
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<tr>
<td>Agenesis of ductus venosus</td>
<td>5</td>
<td>2, cardiomegaly; 3, hydrops; 1, 45,X</td>
<td>4, A&amp;W; 1, IUFD</td>
</tr>
<tr>
<td>Hofstaetter et al¹³</td>
<td>4</td>
<td>3, cardiomegaly; 1, hydrops</td>
<td>1, neonatal death; 1, TOP; 2, alive</td>
</tr>
<tr>
<td>Achiron et al¹⁶</td>
<td>10</td>
<td>2, increased nuchal translucency; 5, cardiomegaly; 2, neonatal death; 5, alive; 1, TOP; 1, IUFD; 1, lost to follow-up</td>
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</tr>
<tr>
<td>Contratti et al³⁸</td>
<td>69</td>
<td>60, without additional findings; 4, transient nuchal findings; 4, minor anomalies; 1, diaphragmatic hernia; 68, healthy after birth</td>
<td></td>
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<tr>
<td>Perimni et al¹⁰</td>
<td>8</td>
<td>7, without additional findings;</td>
<td>Healthy after birth</td>
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<tr>
<td></td>
<td></td>
<td>1, dextrocardia and right-sided descending aorta</td>
<td></td>
</tr>
<tr>
<td><strong>Vitelline veins</strong></td>
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<td></td>
<td></td>
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<tr>
<td>Complete agenesis of portal system</td>
<td>1</td>
<td>Unusual C-shaped vessel between the umbilical vein and dilated inferior vena cava; no portal vein</td>
<td>A&amp;W</td>
</tr>
<tr>
<td>Laverdiere et al⁵⁰</td>
<td>1</td>
<td>Liver hyperechoic foci; dilated inferior vena cava; dilated intraabdominal segment of umbilical vein</td>
<td>A&amp;W</td>
</tr>
<tr>
<td>Venhat-Raman et al⁵⁹</td>
<td>1</td>
<td>Portocaval shunt; no portal vein; right cardiomegaly; partial agenesis of portal system</td>
<td>A&amp;W</td>
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<tr>
<td>Partial agenesis of portal system</td>
<td>1</td>
<td>Umbilical right atrial shunt; liver hyperechoic foci; no ductus venosus; partial agenesis of the portal system</td>
<td>A&amp;W</td>
</tr>
<tr>
<td>Achrom et al¹⁶</td>
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<tr>
<td><strong>Pulmonary Veins</strong></td>
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<td>TAPVCs</td>
<td>4</td>
<td>2, asplenia syndrome; 2, asymmetry of cardiac chambers: right &gt; left</td>
<td>4, neonatal death</td>
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<td>Achiron et al¹⁶</td>
<td>4</td>
<td>2, drainage to the coronary sinus; 1, drainage to the right superior vena cava; 1, drainage to the inferior vena cava; 4, asymmetry of cardiac chambers: right &gt; left</td>
<td>3, neonatal death; 1, alive after successful surgical repair</td>
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<td>Allan and Sharland⁵³</td>
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A&W indicates alive and well; AV, atrioventricular; IUFD, intrauterine fetal death; IUGR, intrauterine growth restriction; LI, left isomerism; NA, not available; PSS, polysplenia syndrome; RI, right isomerism; SI, situs inversus; SIT, situs inversus totalis; and TOP, termination of pregnancy.
Conclusions

On the basis of limited clinical experience and at our present state of knowledge, it is difficult to delineate the fetal and neonatal prognosis when an anomaly of the fetal veins is detected. However, a few conclusions may be drawn from the current published data. Prenatal evaluation of fetuses found to have anomalies of the venous system should include a careful search for cardiac anomalies and a detailed anatomic survey of the umbilical, portal, hepatic, and ductal systems to determine aberrant communication and, if possible, to delineate hints for systemic diseases or a thromboembolic phenomenon. In cases with agenesis of the ductus venosus, careful follow-up is mandatory, because its absence may induce the appearance of hydrops fetalis with an unfavorable prognosis.

References

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