의학석사 학위논문

산전 초음파로 진단된 탯줄문맥전신정맥 단락의 타입에 따른 주산기 예후

Perinatal outcomes according to the type of prenatally diagnosed umbilical-portal-systemic venous shunt

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정 진 하

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이 논문을 의학석사 학위 논문으로 제출함

2021 년 8 월

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국문 요약

서론: 탯줄문맥전신정맥 단락(umbilical-portal-systemic venous shunt, UPSVS)은 매우 드문 선천성 질환으로 탯줄정맥으로부터 간문맥과 전신순환의 연결 이상을 말하며 이로 인하여 간성뇌증이나 폐동맥 고혈압 등의 합병증이 발생할 수 있는 것으로 알려져 있다. UPSVS의 산전진단에 대해 발표된 연구는 있으나 현재까지 타입과 예후에 대한 일관성 있는 결과는 없다. 따라서 본 연구는 산전 진단된 UPSVS 환아들의 초음파 소견과 장기적 예후를 분석하고자 한다.

연구 대상 및 방법: 2005년 1월부터 2020년 9월까지 서울아산병원 산부인과에서 UPSVS를 진단받고 출산한 환자들을 대상으로 후향적 연구를 진행하였다. 본 연구에서는 2016년 Achiron 등에 의해 제시된 분류방법으로 환자를 타입 별로 분류하고 분석하였다. UPSVS를 가진 태아는 타입I(탯줄전신 단락), 타입II(정맥관전신 단락) 및 타입III(문맥전신 단락)으로 분류되며 타입III은 IIIa(간내단락)와 IIIb(간외단락)로 나뉜다. UPSVS의 분류는 산전 초음파 소견과 출생 후 시행한 복부 초음파 및 컴퓨터단층촬영 결과로 타입을 결정하였다. 주산기 예후는 전자의무기록을 이용하여 타입 별로 출생 체중, 심장비대, 동반 기형, 염색체 검사 결과, 이환율 및 사망률을 비교하였다.

결과: 산전에 UPSVS를 진단받은 18명 중, 4명은 산후에 문맥 변이로 확인되어 연구에서 제외하고, 남은 14명을 대상으로 분석하였다. 14명의 진단 당시 임신주수의 중앙값은 30.3주(범위, 23.1-37.3)였고 분만 임신주수의 중앙값은 37.2주(범위, 31.0-39.3)였으며, 출생 체중 중앙값은 2,105g(범위, 1,350-2,870)이었다. 이 중, 11명(79%)은 동반기형이 있었으며

심장비대와 태아수종은 각각 8명(57%)과 2명(14%)에서 관찰되었다. 염색체검사를 시행한 11명 중 3명(27%)에서 염색체 이상이 관찰되었다. 염색체이상으로는 에드워드 증후군, 3번 염색체 단완 결실, 10번과 22번 염색체 불균형전위가 있었다. 사망한 환아는 2명으로, 출생 후 7개월 안에 사망하였으며 모두 염색체 이상을 동반하였다.

가장 흔한 타입은 I 타입(8/14, 57%)이었고, 두번째로 흔한 타입은 IIIa 타입(4/14, 29%)이었다. 나머지 II와 IIIb 타입은 각각 1 명씩 있었다. I 타입은 7명(87%)에서 동반기형이 있었고 5명(63%)에서 심장비대, 5명(63%)에서 태아성장지연을 보였으나 4명(50%)의 환아들은 추적관찰 시기까지 건강하게 생존하고 있었다. IIIa 타입에서는 3명(75%)에서 태아성장지연이 있었으나 출산 후 정상적인 성장을 보이고 있었다. II 타입에 해당하는 1명은 심장비대와 태아 수종, 염색체 이상을 동반하였고 IIIb 타입에 해당하는 1명은 다발성 기형과 염색체이상이 있었다. 환아들의 출생 후 관찰 기간은 3개월에서 최대 6년까지였다. 본 연구의 대상 중 비전형적 문맥 문합으로 인해 수술이나 시술을 받은 증례는 없었다.

결론: UPSVS는 산전에 진단이 가능한 질환으로 산전 초음파 검사상 태아의 복부횡단면에서 제대정맥과 문맥 및 동맥관 연결의 주행이 이상하다면 UPSVS를 의심해야 한다. 또한 산전 초음파 검사에서 UPSVS가 진단되었다면 다른 기형, 염색체 이상, 태아성장지연 또는 심장비대를 동반하고 있지 않은지 확인해야 하며, 다른 동반기형 없이 단독으로 UPSVS만 존재한다면 좋은 예후를 기대할 수 있다. 본 연구 결과는 UPSVS의 주산기 예후에 대한 적절한 상담에 많은 도움을 줄 수 있을 것을

판단된다.

중심단어: 문맥전신 단락, 심장비대, 염색체 이상, 주산기 예후, 태아성장지연, 태아수종, 탯줄문맥전신정맥 단락

차례

표와 그림 목차

Introduction

In a fetal assessment by ultrasonography, an abnormal course of the umbilical vein (UV) at the transverse abdominal view could be indicative of an abnormal fetal venous system. The abnormal course of the UV and the absence or displacement of the portal vein (PV) or ductus venosus (DV) may suggest the presence of other shunts [1]. This condition is rare and has been reported in the context of an absent DV or by using a postnatal dichotomous classification that does not reflect the importance of the DV [2, 3].

In 2016, Achiron *et al.* suggested an in utero classification of umbilical-portal-systemic venous shunt (UPSVS), which include three types: type I, umbilical-systemic shunt (USS); type II, DV-systemic shunt (DVSS); and type III, portal-systemic shunt (PSS). Furthermore, PSS consists of two subtypes: type IIIa, intrahepatic portal-systemic shunt (IHPSS) and type IIIb, extrahepatic porta-systemic shunt (EHPSS) [4]. However, only one study to date has reported the application of this new classification system. In that study, half of the study population was terminated [5]; therefore, it is difficult to describe the characteristics of each UPSVS type based on the available data.

The aim of this study was to report our experience on the prenatal diagnosis of UPSVS according to the in-utero classification discussed and to demonstrate the perinatal outcomes of fetuses according to the type.

Materials and Methods

We retrospectively reviewed fetuses prenatally diagnosed with UPSVS between January 2005 and September 2020 at Asan Medical Center, Seoul, Republic of Korea. This study was approved by the institutional review board of our institution (No. 2021-0277). All prenatal ultrasound examinations underwent an anatomical survey, including fetal echocardiography with Accuvix XQ, V10, A30, or WS80A (Samsung Medison Co., Ltd., Seoul, Republic of Korea), Aloka ProSound α7 or α10 (Aloka Co., Ltd., Tokyo, Japan), or Voluson E8 or E10 Expert (General Electric Healthcare Austria GmbH & Co. OG, Zipf, Austria) equipped with a 2–6 MHz transabdominal transducer. We included patients based on the following inclusion criteria: prenatally diagnosed with UPSVS, delivered at our institution, postnatally confirmed to have UPSVS, and followed up for at least one month after birth. We excluded patients who were lost to follow-up.

Analysis of prenatal ultrasonographic findings

We evaluated the transverse abdominal view to confirm the integrity of the umbilical-portal-DV complex and classified the cases according to UPSVS type. The UPSVS types are described in Table 1, and schematic diagrams of each type are shown in Figure 1. One expert in maternal-fetal medicine reviewed all ultrasonographic images with medical reports and classified the cases according to UPSVS type.

Analysis of clinical features

Prenatal data included maternal age, gestational age (GA) at diagnosis, chromosome analysis, associated anomaly, fetal hydrops, fetal cardiomegaly, fetal growth restriction, type of UPSVS, GA at delivery, and birth weight. Fetal growth restriction was defined as birth weight below the 10^{th} percentile based on Asian standards for fetal growth [6]. Fetal cardiomegaly was defined as a cardiothoracic (C/T) area ratio or a C/T circumference ratio greater than two standard deviations [7]. Fetal hydrops is a fetal condition defined as abnormal accumulation of fluid in two or more spaces, which may include ascites, pleural effusion, pericardial effusion, and skin edema.

All babies were examined by neonatal specialists and had postnatal diagnosis of UPSVS, which was further confirmed by abdominal ultrasound or computed tomographic (CT) angiography. Postnatal data included clinical and biochemical findings caused by UPSVS, any indication for surgery or intervention for UPSVS, and duration of follow-up. Structural abnormalities found after birth were also included.

Statistical analysis

The data were presented as medians with ranges or as numbers with percentages.

Results

Among the 18 patients prenatally suspected with UPSVS, 14 were confirmed with UPSVS postnatally and four were confirmed with PV variants. The most common type of USPVS was type I (n=8, 57%), followed by type IIIa (n=4, 29%), type II (n=1, 7%), and type IIIb (n=1, 7%). The clinical characteristics of the study population are summarized in Table 2. Cardiomegaly and fetal hydrops were observed in eight (57%) and two (14%) patients, respectively. Among the 11 patients who underwent chromosomal analysis, three had abnormal results: trisomy 18, 3p deletion syndrome, and unbalanced (X;22) translocation. Two of the patients died during the infant period. Table 3 presents the detailed outcomes of all the patients diagnosed with UPSVS.

Type I, USS

There were eight cases of type I UPSVS. These cases did not show any DV, and the UV was connected to the systemic vein. In six cases (cases 1, 4–8), the UV directly drained into the right atrium (Figure 2). In two cases (cases 2 and 3), the UV drained into the inferior vena cava (IVC). All but one (7/8, 88%) were associated with other structural abnormalities: two cases with VACTERL association (cases 6 and 8) and one case with Cornelia de Lange syndrome (case 3), which shows distinctive facial characteristics and growth delays [8]. The patient with trisomy 18 died at the age of seven months because of respiratory failure due to tracheomalacia (case 5). Cardiomegaly was observed in five fetuses, three of which resolved. The median GA

at birth was 38.1 weeks (range, 33.3–39.4) and the median birth weight was 2,540 g (range, 1,910–2,870). Five neonates had birth weights below the 10th percentile, and four of them were below the 3rd percentile.

Type II, DVSS

We found one case of type II UPSVS (case 9). The fetus had a short DV that drained into the IVC, but other structures of the umbilical-portal-DV complex were intact. The fetus showed cardiomegaly and fetal hydrops with severe skin edema and pleural effusion (Figure 3). This fetus was confirmed to have a 3p deletion. The fetal hydrops resolved after birth. At the time of writing, the patient was 17 months old and had developmental delay.

Type IIIa, IHPSS

Four cases were classified as type IIIa UPSVS. They showed an abnormal connection between the PV and hepatic vein, which was observed as a tortuous and engorged vessel in the fetal liver (Figure 4). However, the umbilical-portal-DV complex remained intact. Among these cases, three (75%) had birth weights below the 10th percentile and all had transient mild hyperammonemia. More importantly, none of them required shunt ligation surgery or liver transplantation.

Type IIIb, EHPSS

One case of type IIIb UPSVS was identified (case 14). Prenatal ultrasonography revealed

an abnormal tortuous vessel communicating between the PV and IVC (Figure 5). Due to this shunting flow, the IVC became enlarged; otherwise, the umbilical-portal-DV complex was intact. The fetus was below the 3rd percentile and had both cardiac and extra-cardiac anomalies. Additionally, the karyotype results showed an unbalanced (X;22) translocation. The neonate suffered from bronchial stenosis and died due to pulmonary hypertension and heart failure at the age of three months.

PV variant group

Four cases were confirmed with PV variants on postnatal abdominal sonography. In the prenatal period, the connection between the left PV and the UV was not clear. However, the DV was observed and there was no evidence of PSS. Thus, we could not classify these cases as any type of UPSVS. Furthermore, postnatal ultrasonography showed a PV variant, which was a left PV arising from a right PV or a main PV. Although all patients had associated anomalies, there were no cases of cardiomegaly, and there was one case of growth restriction (Table 4).

Discussion

According to our study, prenatal diagnosis and classification of UPSVS are feasible. We introduced 14 cases of UPSVS based on the new classification. These cases were mainly associated with growth restriction and structural abnormalities, and some were accompanied by abnormal chromosomal results. In patients with type I, growth restriction and cardiomegaly are common. If these patients have no other structural abnormalities, the fetus can live well. Nevertheless, accompanying structural anomalies and chromosomal abnormalities may still appear for some; in such cases, targeted ultrasound and chromosomal studies should be recommended.

The previous classification was used without considering the DV. On the other hand, the new in utero UPSVS classification system was based on the embryological-anatomical origin of the shunt. As such, abnormalities in the fetal venous system can be diagnosed by characterizing the prenatal blood flow system.

In South Korea, a study on congenital portosystemic shunts was reported in 2013 [9]. The authors categorized the cases according to the classification suggested by Park *et al.* in 1990 [10]. Among the six cases included, shunts were diagnosed prenatally in only two cases. The four other cases showed a prominent hepatic vein, abnormal intrahepatic tubular structure, cardiomegaly, or intrauterine growth restriction. Compared to this study, our study shows that all UPSVS cases were diagnosed before birth and were categorized using the new

classification of UPSVS discussed.

Abnormalities associated with the fetal venous system are relevant to fetal growth restriction. In previous reports, the overall percentage of intrauterine growth retardation observed in patients with UPSVS was 39.5%, with more than 50% reported as types I and IIIa [3]. In this study, 64% of the cases were associated with fetal growth restriction. When comparing the frequency of growth restriction by UPSVS types, type IIIa was the most common with 75% (3/4), which was followed by type I with 63% (5/8). Overall, 55% (5/9) showed catch-up growth after birth. Since there were many cases associated with multiple anomalies, the catch-up growth rate was lower compared to most small for gestational age children. The fetus without major anomalies can grow well after birth due to changes in the portal system. According to a previous study, severe fetal growth restriction resulted in increased DV flow and reduced umbilical vein flow to the liver. Decreased hepatic flow may induce fetal growth restriction as a result [11]. They suggested that the diameter of the right portal vein could be a reliable ultrasound marker to predict fetal growth restriction. Since the UV drains directly into the systemic blood flow in type I UPSVS, there is decreased blood flow in the liver. Consequently, growth restriction may occur in type I UPSVS. Likewise, PSS reduces blood flow to the liver parenchyma, affecting the growth process.

Cardiomegaly was observed in eight fetuses (57%). It resolved in four of these fetuses before birth and in three infants at one week, 12 months, and 31 months after birth, respectively.

At the time of writing, the remaining patient still had right ventricular hypertrophy due to associated cardiac anomalies. In the absence or disposition of the DV, cardiomegaly can occur because of the dysregulation of venous return to the heart [12]. In this study, in the absence of congenital heart disease, cardiomegaly spontaneously improved in most cases.

Previous studies have reported that type I UPSVS has the poorest prognosis with the lowest rates of live birth and postnatal survival [4, 5]. In these two studies, 10 out of 13 cases were terminated due to complete absence of an IHPVS. Therefore, these studies were limited in their ability to evaluate perinatal outcomes. In the present study, half of the type I cases were alive and doing well without major anomalies at the time of writing.

There were three cases of chromosomal abnormalities observed in this study that were not reported in any previous studies: trisomy 18, 3p deletion, and unbalanced (X;22) translocation. Several chromosomal anomalies have been reported in cases of abnormal fetal venous systems. Among these, trisomy 21 was the most common karyotype anomaly and was mainly associated with type II UPSVS. Before the introduction of in-utero classification, a study on 19 fetuses with absent DV and umbilical venous drainage in the portal sinus reported two cases of 13p deletion and 4p deletion. The chromosomal abnormalities observed in the three cases in this study were characterized by multiple structural anomalies. Trisomy 18 is characterized by an abnormal central nervous system, congenital heart disease, and limb anomaly [13]. Microcephaly and congenital heart disease were found in the 3p deletion. Unbalanced (X;22)

translocation could have similar features as DiGeorge syndrome [14]. There could be UPSVS in these diseases showing multiple anomalies, but it may not have been detected previously due to low level of awareness about UPSVS.

In this study, four cases of PV variation were identified. In the transverse abdominal view, UV insertion and connection of the PV were intact, but the left PV was not identified. The DV was found to be intact, and another right PV course was also observed. Based on our experience, PV variants can be prenatally mistaken for UPSVS. Since the PV was too thin to be evaluated even after birth, it took several months to diagnose the PV variant. We found that an intact umbilical-portal-DV complex and the absence of a portal-systemic shunt could be the points of differential diagnosis between UPSVS and PV variants. Although they had combined anomalies, all four cases were alive at the time of writing. Considering these, PV variation may not be clinically significant in the absence of liver disease.

In conclusion, although UPSVS is a rare fetal disease, prenatal diagnosis of UPSVS is feasible. When an abnormal course of the UV or PV is detected on the transverse abdominal view, UPSVS should be suspected. According to our study, type I and IIIa UPSVS were common. Furthermore, UPSVS is commonly associated with fetal growth restriction, cardiomegaly, structural anomalies, and chromosomal anomalies. UPSVS without other conditions may notably have favorable prognosis. This study was limited by the inability to evaluate the perinatal outcomes of all types due to the small number of cases. Nevertheless,

our study introduced additional information about UPSVS and can both help predict the prognosis of UPSVS and assist in prenatal counseling.

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Table 1. In-utero classification of UPSVS

Type	Name	Description
I	USS	Agenesis of both the DV and the left branch of portal vein results in a normal IHPVS failing to form. Umbilical vein is directly connected to the systemic circulation.
II	DVSS	A short DV is connected to the systemic vein with an intact umbilical-portal-DV complex. Mainly, the location of DV is different or shorter than usual.
III	PSS	
IIIa	IHPSS	The shunt between IHPVS and the hepatic veins is present.
IIIb	EHPSS	The shunt between the portal system and systemic veins is present.

UPSVS, umbilical-portal-systemic venous shunt; USS, umbilical-systemic shunt; DV, ductus venosus; IHPVS, intrahepatic portal venous system; DVSS, ductus venosus-systemic shunt; PSS, portal-systemic shunt; IHPSS, intrahepatic portal-systemic shunt; EHPSS, extrahepatic portal-systemic shunt.

Table 2. Clinical characteristics of UPSVS (N=14)

Characteristics	
Maternal age (years)	35.5 (28–40)
GA at diagnosis (weeks)	30.2 (23.1–37.3)
GA at birth (weeks)	37.2 (31.0–39.3)
Birth weight (g)	2,105 (1,350–2,870)
<10th percentile	9/14 (64%)
Abnormal karyotype (n, %)	3/11 (27%)
Trisomy 18 3p deletion Unbalanced (X;22) translocation	1 1 1
Overall survival rate at 28 days after birth	100%
Associated anomaly (n, %)	11/14 (79%)

UPSVS, umbilical-portal-systemic venous shunt; GA, gestational age.

Data are presented as median (range) or number (%).

Table 3. Detailed clinical outcomes of 14 cases of UPSVS

Case	UPSVS type	Associated anomaly	Karyotype	Fetal hydrops	Cardiomegaly	Growth restriction	GAD (weeks)	BW (g)	Follow-up period
1	I	minor	Normal	-	+	-	38.0	2,800	3 m
2	I	None	Normal	-	+	+*	38.3	2,040	6 y
3	I	Multiple anomalies	Normal	+	-	+*	37.1	1,980	2 m
4	I	Isolated extracardiac	-	-	+	-	39.1	2,870	4 y
5	I	Multiple anomalies	Trisomy 18	-	-	+	38.6	2,580	Died at 7 m
6	I	Multiple anomalies	Normal	-	-	+*	37.6	1,910	2 y 7 m
7	I	Isolated cardiac	Normal	-	+	-	33.3	2,560	2 y 7 m
8	I	Multiple anomalies	Normal	-	+	+*	39.4	2,520	1 y 6 m
9	II	Multiple anomalies	3p deletion	+	+	-	31.0	2,170	1 y 5 m
10	IIIa	Isolated cardiac	Normal	-	+	+*	36.6	1,793	1 y 2 m
11	IIIa	None	-	-	-	-	36.6	2,490	2 y 3 m
12	IIIa	None	Normal	-	-	+*	34.6	1,550	7 m
13	IIIa	Minor	-	-	+	+*	37.4	1,900	4 m
14	IIIb	Isolated cardiac	t(X;22)	-	-	+*	32.4	1,350	Died at 3 m

UPSVS, umbilical-portal-systemic venous shunt; GAD, gestational age at delivery; BW, birth weight; t(X;22), unbalanced (X;22) translocation; m, months; y, years

* below 3rd percentile birth weight.

Table 4. Detailed clinical outcomes in 4 cases of portal vein variant

Case	Associated anomaly	Karyotype	Fetal hydrops	Cardiomegaly	Growth restriction	GAD (weeks)	BW (g)	Follow-up period
15	Isolated extracardiac	Normal	-	-	+	38.3	2,067	7 m
16	Isolated cardiac	Normal	-	-	-	40.0	3,230	4 y 2 m
17	Multiple anomalies	Normal	-	-	-	35.4	2,800	2 y 5 m
18	Multiple anomalies	Normal	-	-	-	38.0	3,010	11 m

GAD, gestational age at delivery; BW, birth weight; m, months; y, years.

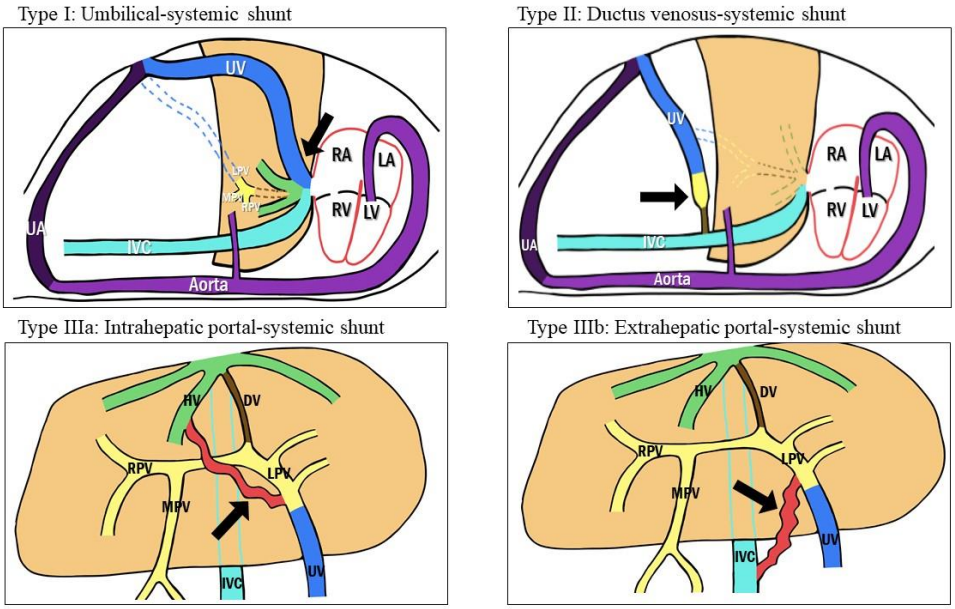

Figure 1. Schematic diagrams of umbilical-portal-systemic venous shunt. Shunts are indicated as arrows. UA, umbilical artery; UV, umbilical vein; IVC, inferior vena cava; RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; LPV, left portal vein; MPV, main portal vein; RPV, right portal vein; HV, hepatic vein; DV, ductus venosus.

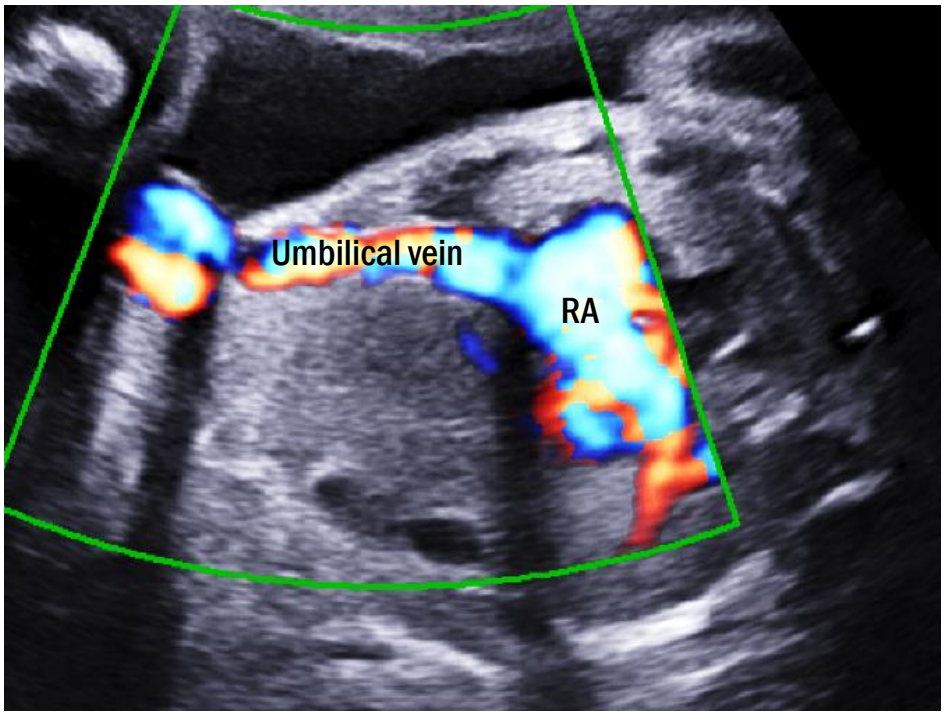

Figure 2. Two-dimensional color Doppler image of type I umbilical-portal-systemic venous shunt in case 7. Umbilical vein directly drains into the right atrium without going through the ductus venosus. RA, right atrium.

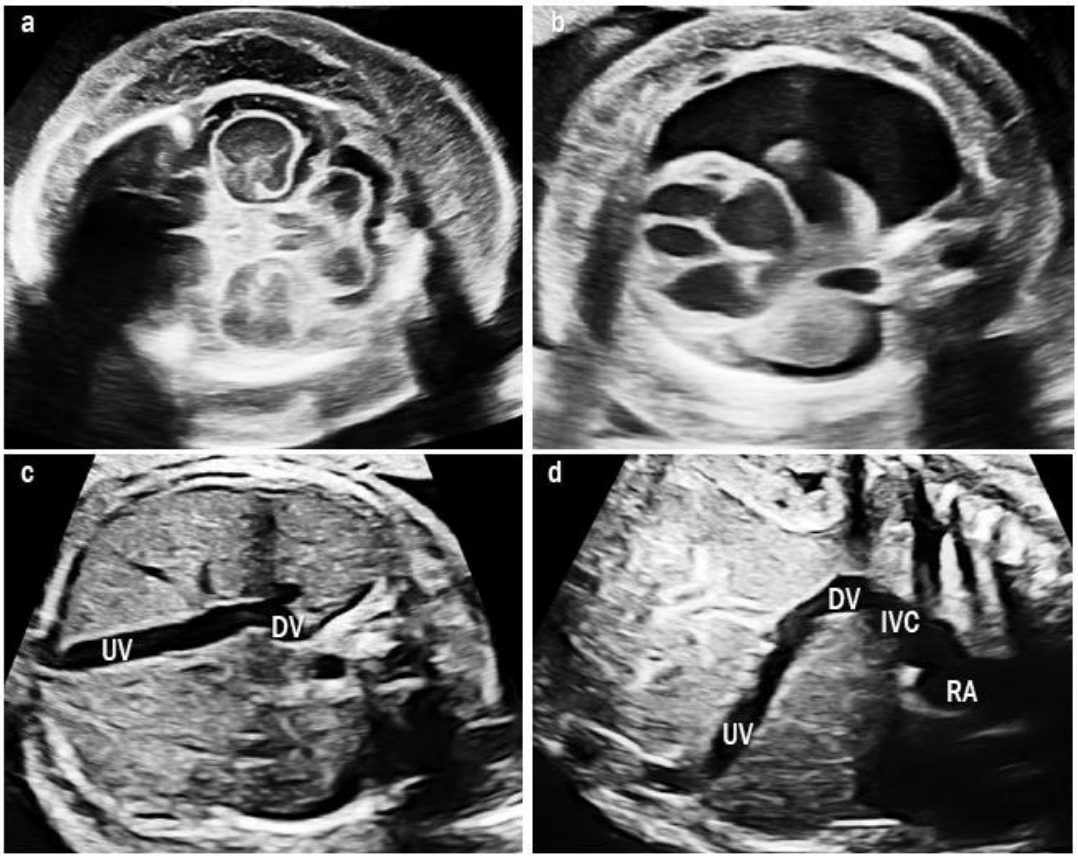

Figure 3. Ultrasound images of type II umbilical-portal-systemic venous shunt in case 9. The fetus showed severe fetal hydrops: (a) severe skin edema and (b) pleural effusion. (c) Transverse abdominal image shows the UV connected to the DV and it drains at the lower site than usual. (d) Sagittal image shows a short DV draining into the IVC below the diaphragm. UV, umbilical vein; DV, ductus venosus; IVC, inferior vena cava; RA, right atrium.

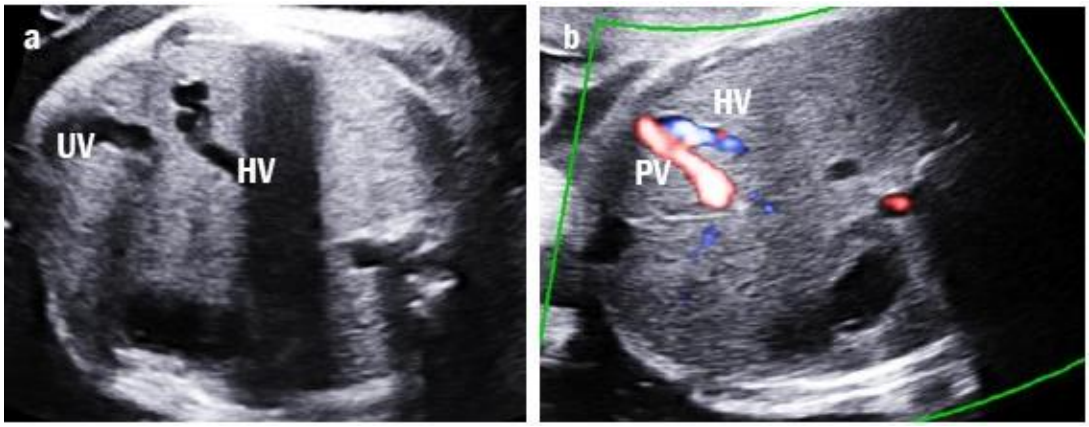

Figure 4. Intrahepatic portal-systemic shunt in case 11. (a) The tortuous and engorged hepatic vein (HV) is shown. (b) The intrahepatic portal vein (PV) is connected to the HV.

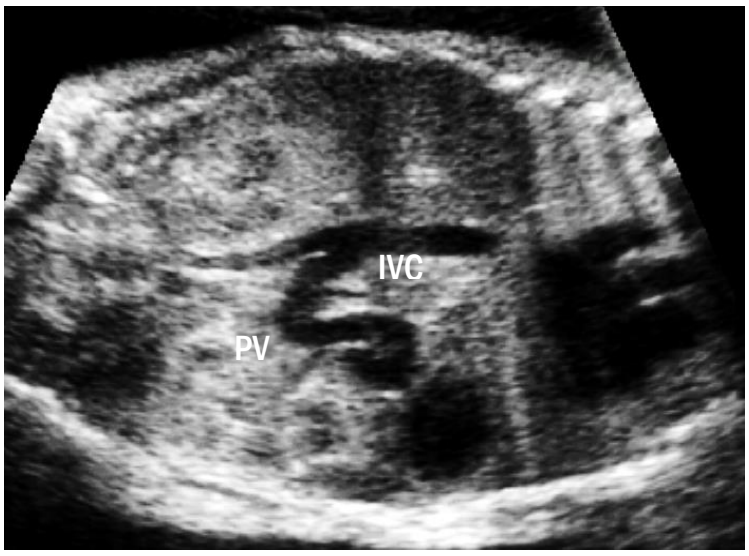

Figure 5. Extrahepatic portal-systemic shunt in case 14. The portal vein (PV) is connected to the inferior vena cava (IVC).

영문 요약

Objective: To report our experience with prenatal diagnosis of umbilical-portal-systemic venous shunts (UPSVS) and to evaluate the perinatal outcomes of fetuses with different types of UPSVS.

Methods: This was a retrospective study of fetuses prenatally diagnosed with UPSVS between January 2005 and September 2020 at Asan Medical Center. UPSVS was prenatally classified into three types according to the anatomical origin of the shunt: type I, umbilical-systemic shunt; type II, ductus venosus-systemic shunt; and type III, portal-systemic shunt. Type III was further divided into two subtypes: type IIIa, intrahepatic shunt; type IIIb, extrahepatic shunt. Postnatal ultrasonography or abdominal computed tomography (CT) was performed to confirm UPSVS.

Results: Out of 18 fetuses prenatally diagnosed with UPSVS, four were excluded. All four patients were confirmed to have portal vein variations without a shunt. The median gestational age at the diagnosis of the remaining 14 fetuses was 30.3 weeks (range, 23.1–37.3). The median gestational age at birth was 37.2 weeks (range, 31.0–39.3), and the median birth weight was 2,105 g (range, 1,350–2,870), including nine (64%) who had body weights below the 10th percentile. Eleven patients (79%) had combined anomalies; cardiomegaly and fetal hydrops were observed in eight (57%) and two (14%) patients, respectively. Among the 11 patients who underwent chromosomal analysis, three had abnormal results. Two infants with abnormal

karyotypes died. The most common type was type I (8, 57%), followed by type IIIa (4, 29%), type II (1, 7%), and type IIIb (1, 7%). In type I, all but one patient had structural anomalies; five (63%) showed cardiomegaly and five (63%) were associated with growth restriction. Nevertheless, half of the patients survived without major anomalies. In type IIIa, three (75%) patients were associated with growth restriction, but they showed catch-up growth after birth. All patients had mild transient hyperammonemia. One type II patient was associated with cardiomegaly, fetal hydrops, and an abnormal karyotype. One type IIIb patient had multiple anomalies and an abnormal karyotype. No surgical correction was required because of the UPSVS.

Conclusion: Prenatal diagnosis of UPSVS is feasible. When an abnormal course of the umbilical or portal vein is detected on the transverse abdominal view, UPSVS should be suspected. UPSVS is commonly associated with structural anomalies, chromosomal anomalies, growth restriction, and cardiomegaly. UPSVS without associated anomalies may have a favorable prognosis. Our study can help predict the prognosis of UPSVS and assist in prenatal counseling.

Keywords: cardiomegaly, chromosome disorders, fetal growth restriction, perinatal outcome, portal vein, portasystemic shunt