CERPO

Centro de Referencia Perinatal Oriente

Facultad de Medicina, Universidad de Chile

AGENESIA DE DUCTUS VENOSO

Dra. Ana Luisa Perez Mendez

Programa Especialización Medicina Materno Fetal

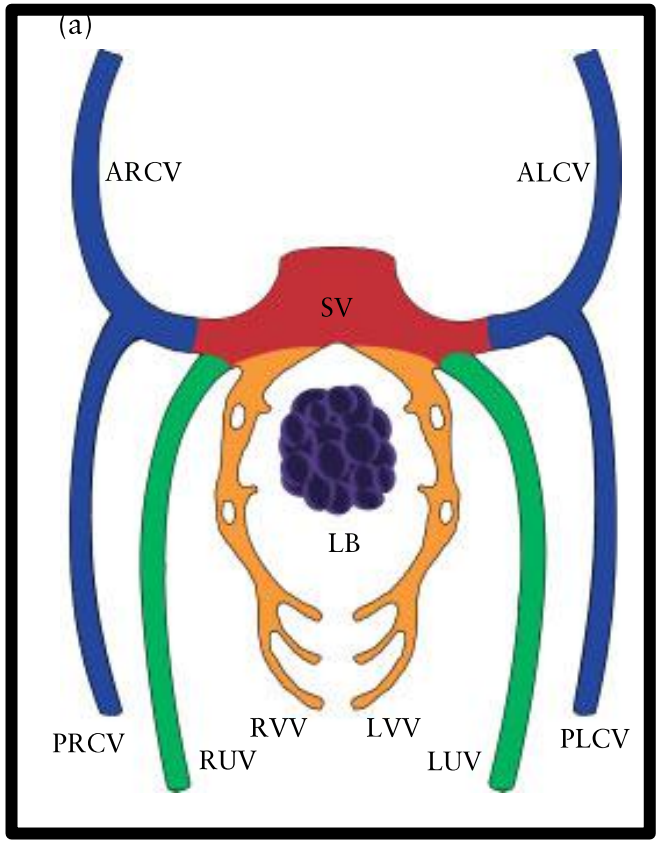
SISTEMA VENOSO FETAL

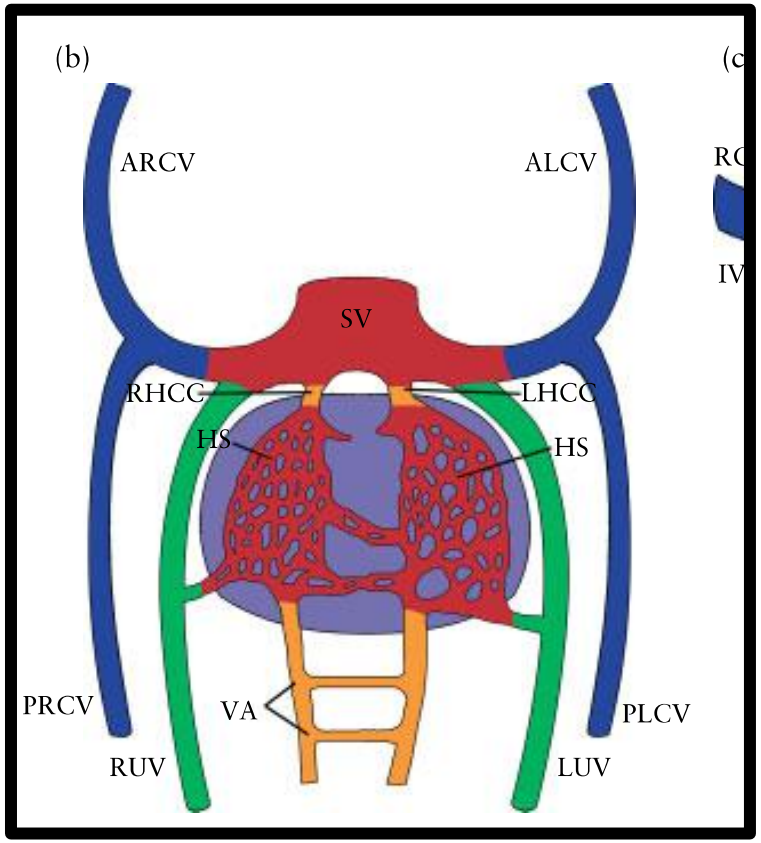
- EL SISTEMA CARDIOVASCULAR FETAL ES EL PRIMERO EN DESARROLARSE EN EL EMBRIÓN HUMANO
- TRES PARES DE VENAS SIMETRICAS FORMAN LA BASE DEL SISTEMA VENOSO PRIMITIVO

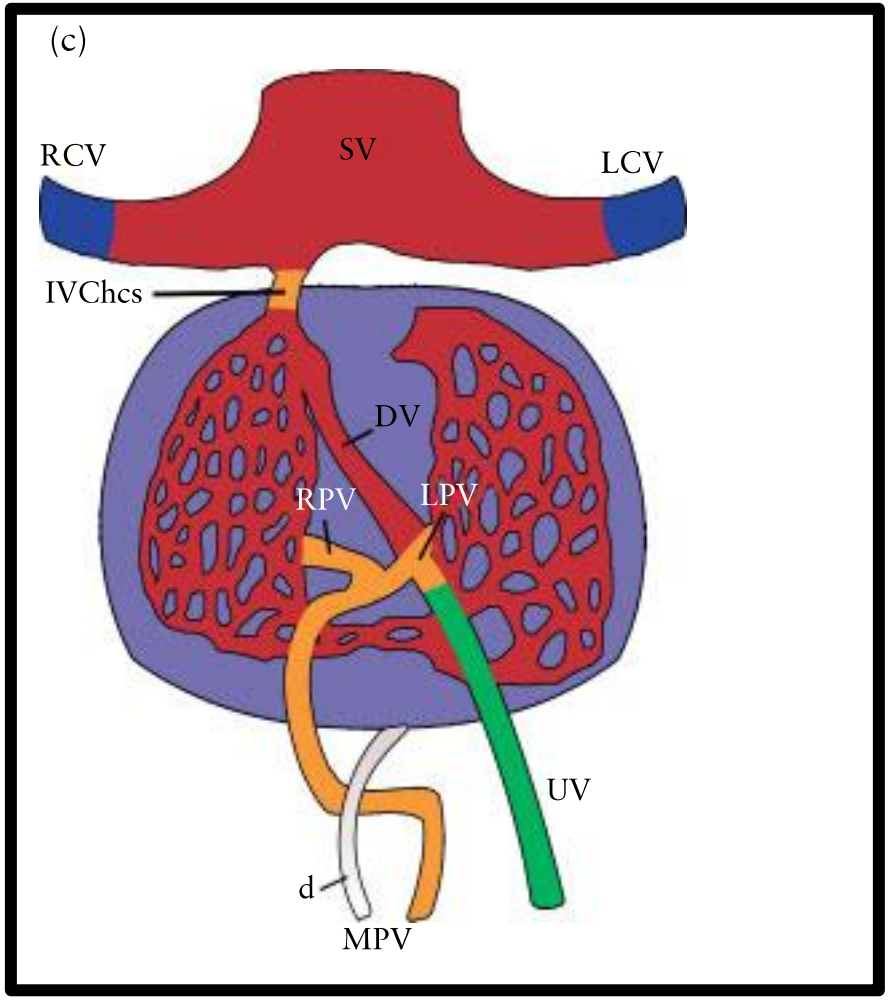
VENAS UMBILICALES (UVS) CORION

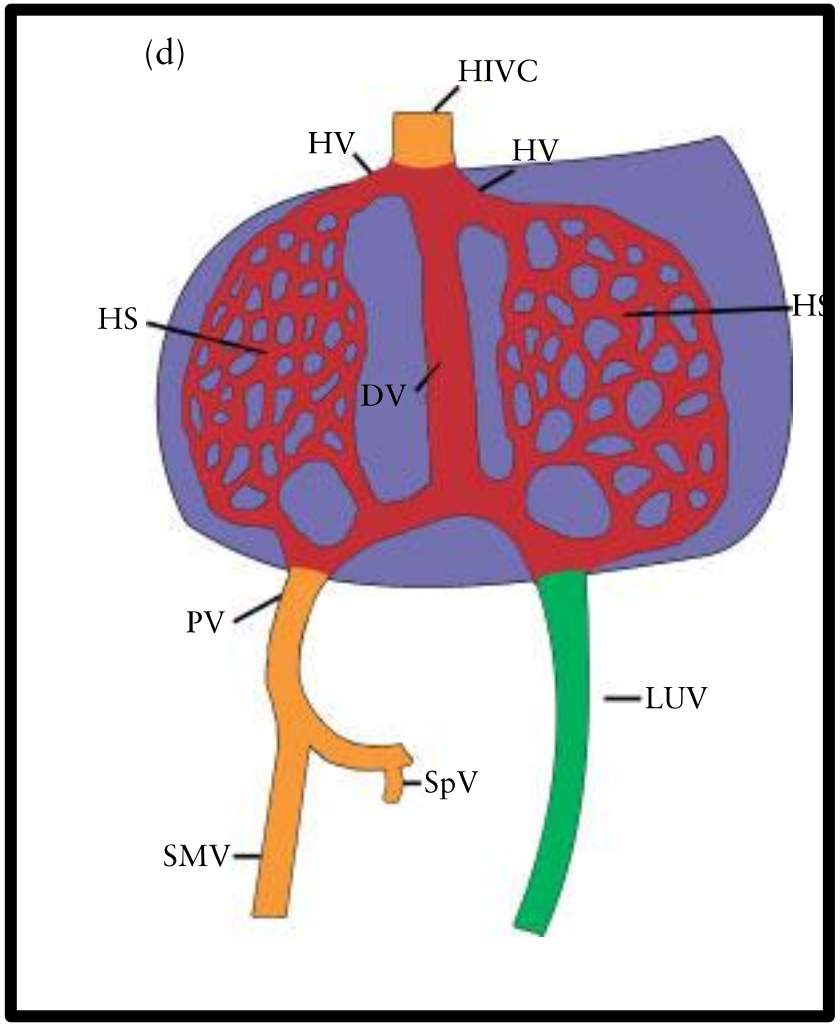
VENAS VITELINAS (VVS) SACO VITELINO

VENAS CARDINALES (CVS) CUERPO DEL EMBRION

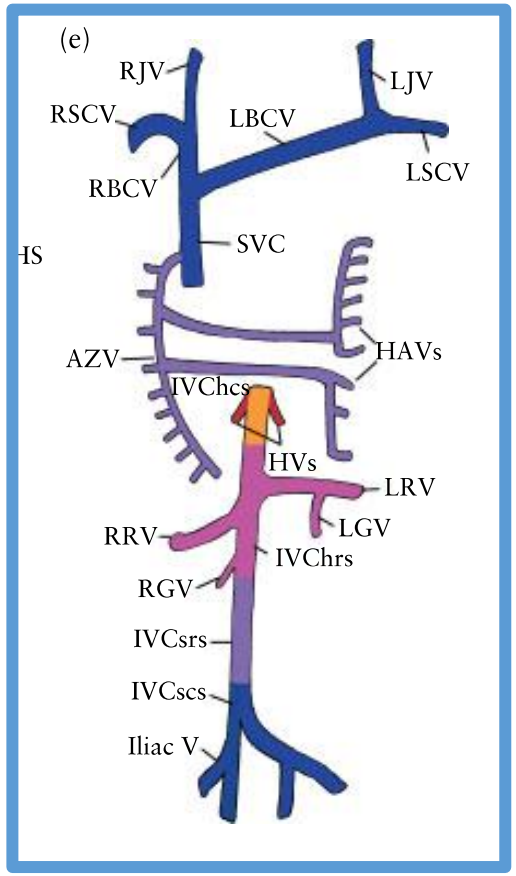

(a)
ARCV
ALCV
SV
LB
PRCV
RUV
RVV
LVV
LUV
PLCV


(b)
(c
ARCV
ALCV
RC
IV
SV
RHCC
LHCC
HS
HS
PRCV
VA
PLCV
RUV
LUV

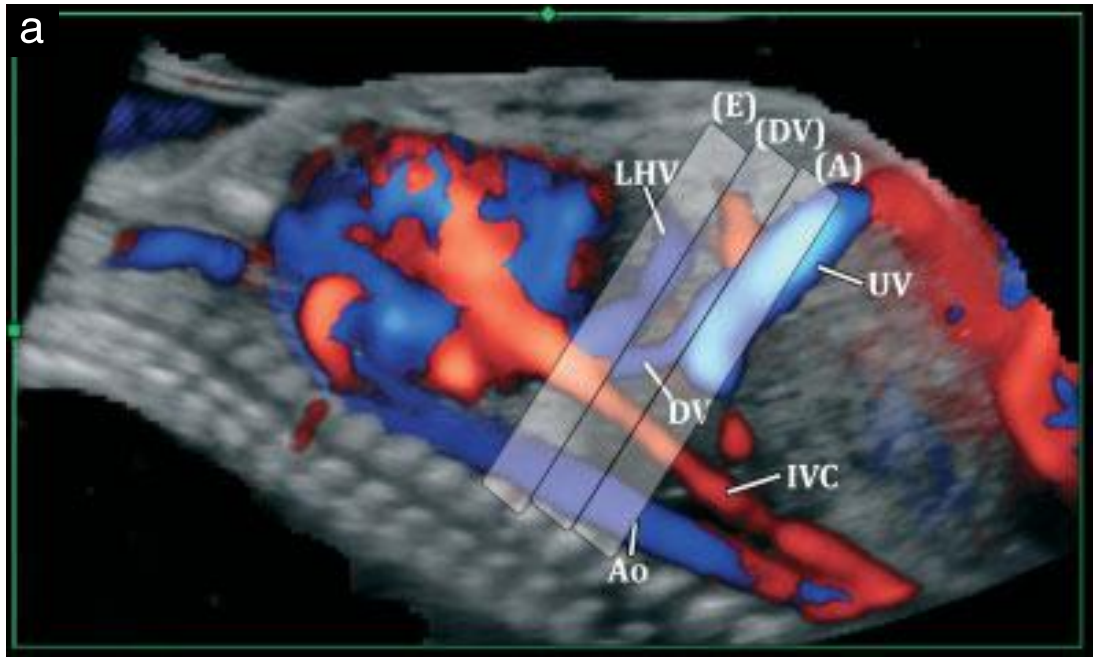

(c)
RCV
SV
LCV
IVChcs
DV
RPV
LPV
UV
d
MPV


(d)
HIVC
HV
HV
HS
DV
PV
LUV
SpV
SMV

CLASIFICACION DE LAS ANOMALIAS DEL SISTEMA VENOSO



- **A)VENAS CARDINALES**
  - **SD HETEROTAXIA**
  - **VCSP,VCIIP**
- **B)VENAS UMBILICALES:**
  - **FALLAS DE LA ANASTOMOSIS CON AGENESIA DE DUCTUS VENOSO**
  - **VENA UMBILICAL DERECHA PERSISTENTE**
- **C)VENAS VITELINAS**
  - **SHUNT PORTOSISTEMICO**
- **D)ANOMALIAS DEL SISTEMA VENOSO PULMONAR**
  - **DRENAJE VENOSO ANOMALO**


a
(E)
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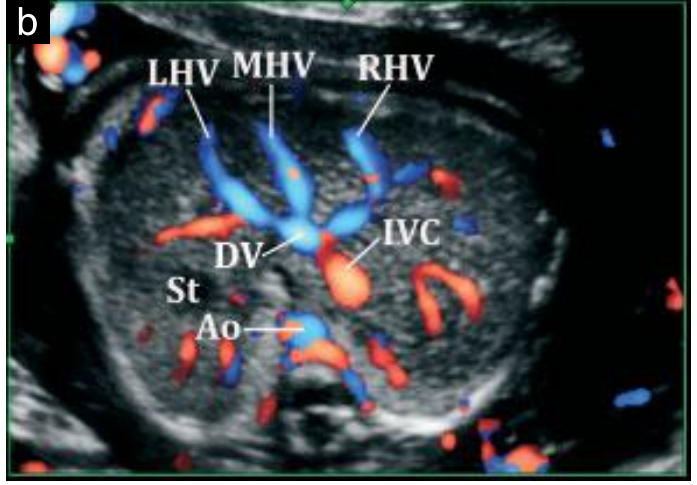
(A)
LHV
UV
DV
IVC
Ao


b
LHV
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c
DV
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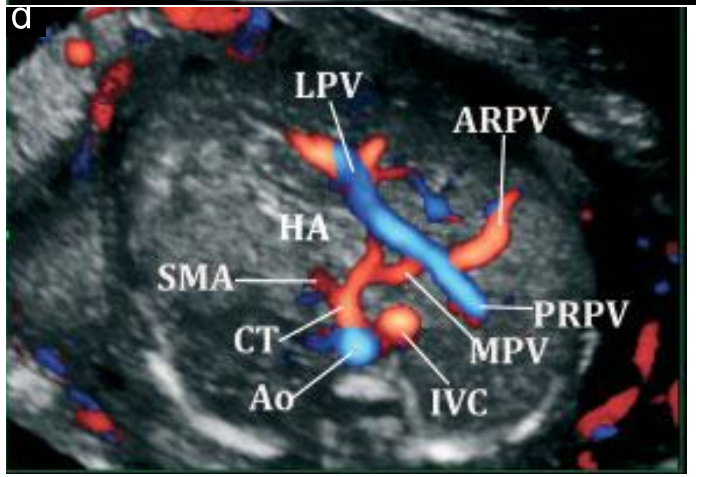
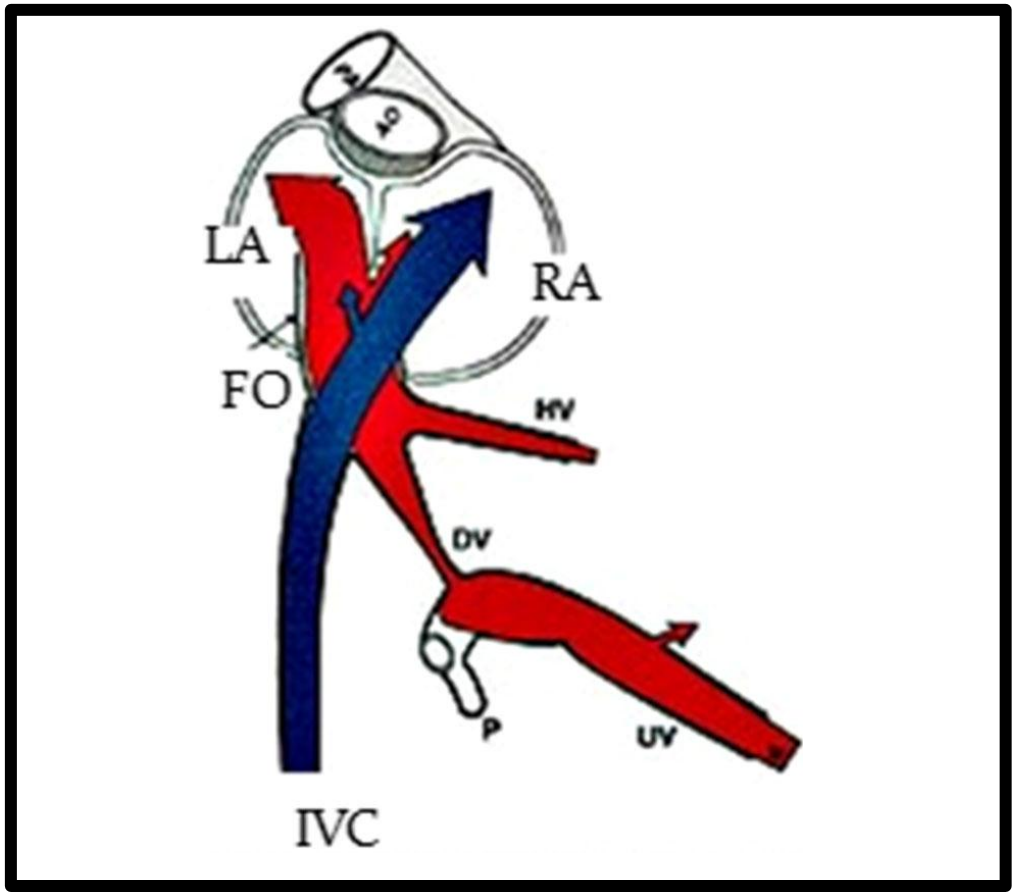
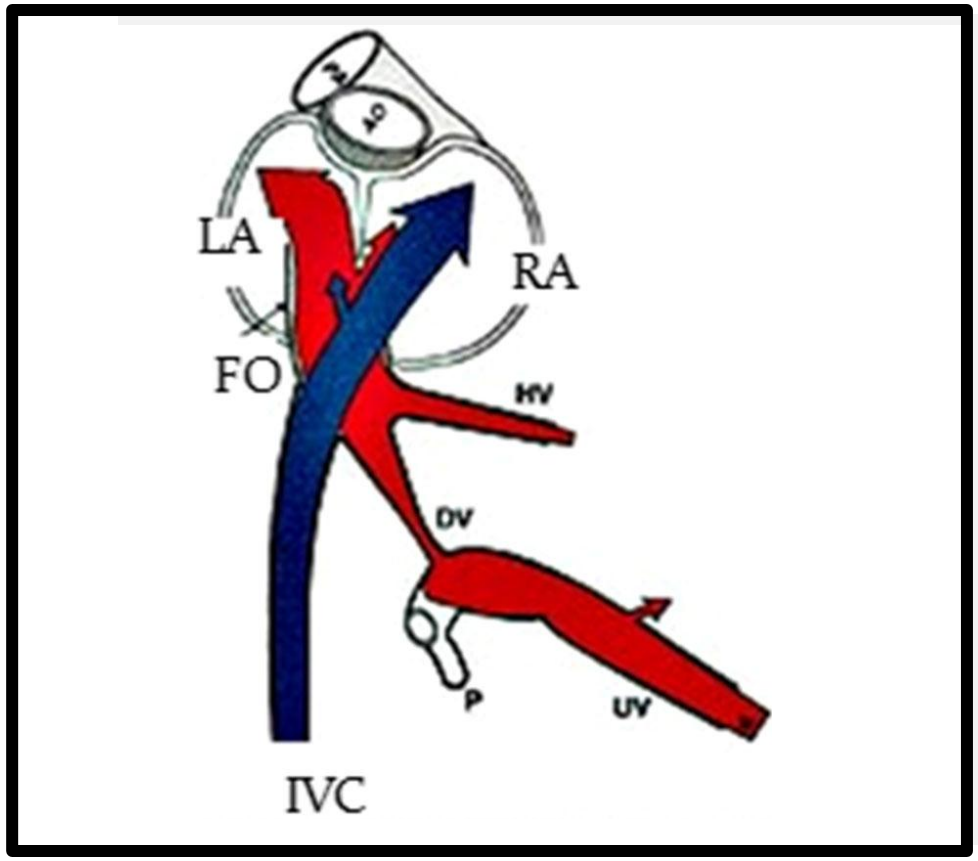

d
LPV
ARPV
HA
SMA
PRPV
CT
MPV
Ao
IVC

Ductus Venoso



- LA SANGRE OXIGENADA PROVENINETE DE LA PLACENTA ALCANZA EL CORAZÓN FETAL MEDIANTE LA VENA UMBILICAL A TRAVES DEL DUCTUS VENOSO

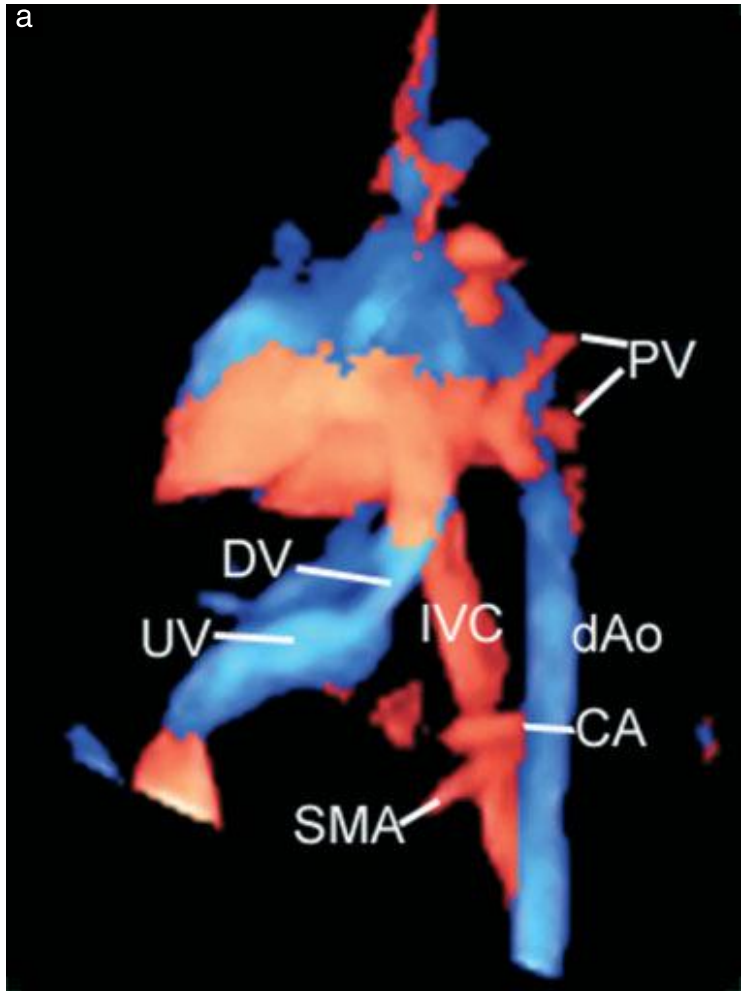


FLUJO UMBILICAL FETAL AUMENTA CON LA EDAD GESTACIONAL
33-54 ML /MIN 20-23 SEM
221-320/ML MIN 36-38 SEM

PERO EL CRECIMIENTO FETAL ES ACELERADO Y ESTO ES INVERSO RESPECTO A SUPERFICIE CORPORAL

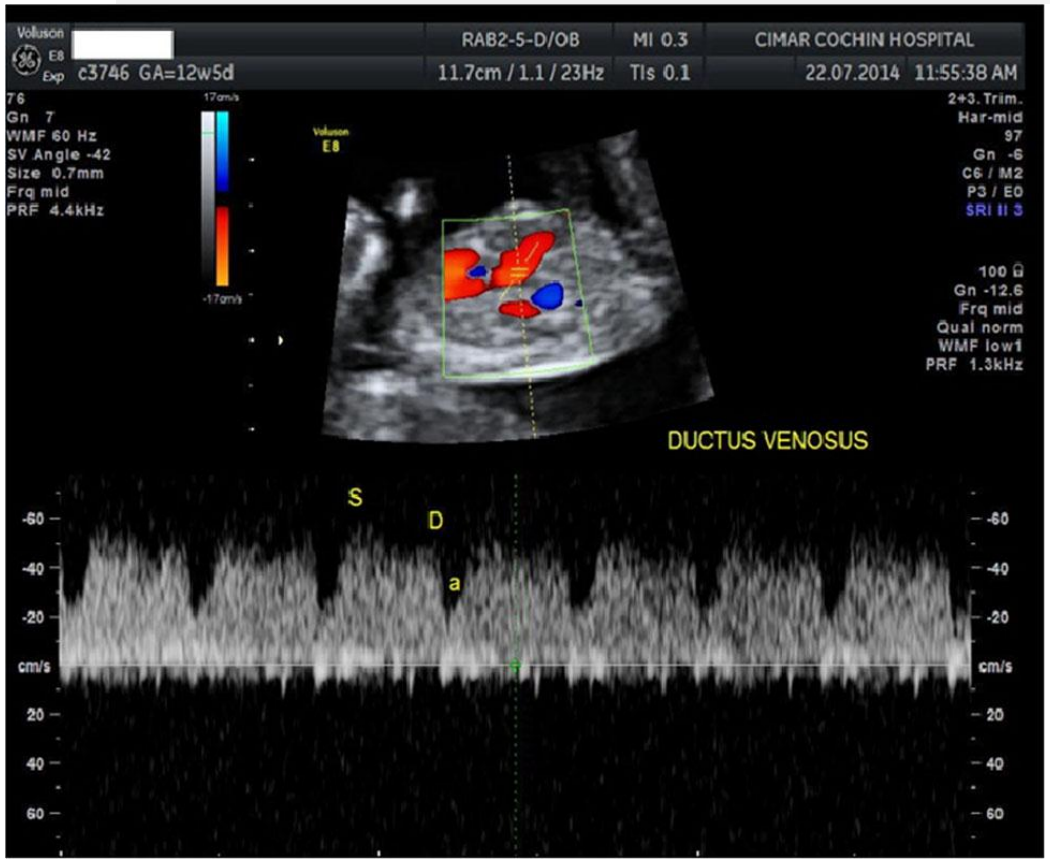
117- 125 ML MIN/KG 20-23
63-104 ML MIN/KG 36-38 SEM

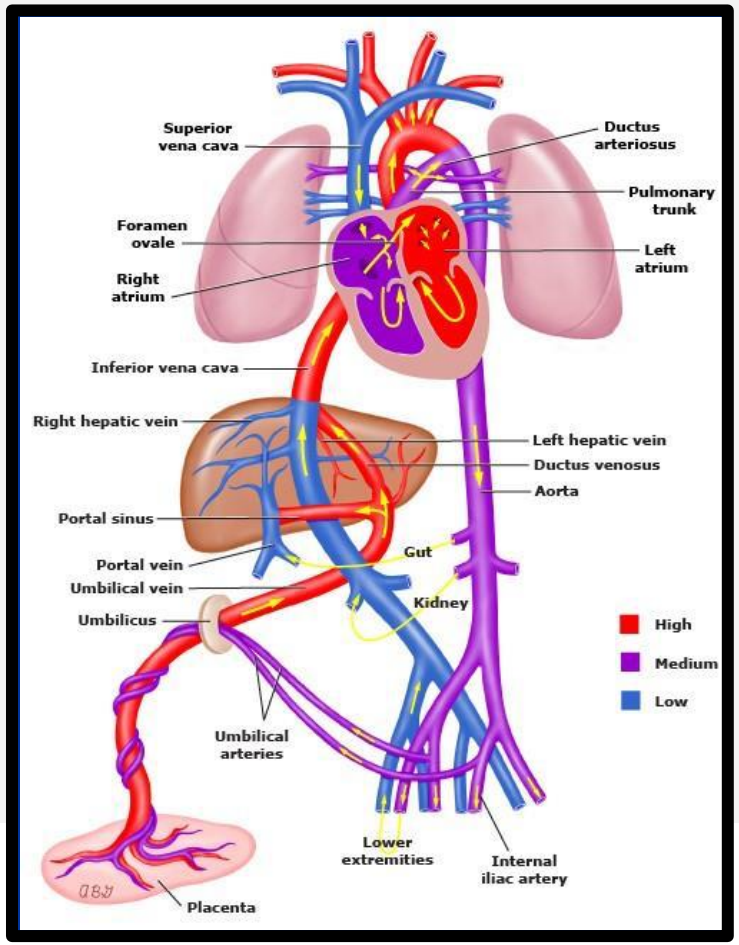
- EL DUCTUS VENOSO ES UN VASO EN FORMA DE RELOJ DE ARENA DE 1 A 2 MM DE ANCHO, APROXIMADAMENTE UN TERCIO DE LA VENA UMBILICAL
- SISTEMA “ESFINTER” DEPENDIENTE DE LA CONCENTRACION DE OXIGENO
- HISTOLOGICAMENTE NO EXISTE TAL ESFINTER



- GRADIENTE PRESION UMBILICALCAVA
- SINERGIA ENTRE CONTRACTILIDAD MIOCARDICA Y BAJA RESISTENCIA PLACENTARIA

- AUMENTO DE GRADIENTE MEDIANTE DUCTUS VENOSO
- ACELERACIÓN DE 15-17 CM/ SEG EN VENA UMBILICAL A 65-75 CM SEG EN DUCTUS VENOSO
- UN 20%-30% DE LA SANGRE OXIGENADA DE LA VENA UMBILICAL LLEGA DIRECTAMENTE A A LA AD GRACIAS AL DV


Voluson E8 Exp
c3746 GA=12w5d
RAB2-5-D/OB
MI 0.3
11.7cm / 1.1 / 23Hz
TIs 0.1
CIMAR COCHIN HOSPITAL
22.07.2014 11:55:38 AM
76
Gn 7
WMF 60 Hz
SV Angle -42
Size 0.7mm
Frq mid
PRF 4.4kHz
17cm/s
-17cm/s
2+3. Trim.
Har-mid
97
Gn -6
C6 / M2
P3 / E0
SRI II 3
100
Gn -12.6
Frq mid
Qual norm
WMF low1
PRF 1.3kHz
DUCTUS VENOSUS
S
D
a
-60
-40
-20
cm/s
20
40
60


Superior vena cava
Ductus arteriosus
Pulmonary trunk
Foramen ovale
Left atrium
Right atrium
Inferior vena cava
Right hepatic vein
Left hepatic vein
Ductus venosus
Aorta
Portal sinus
Gut
Portal vein
Umbilical vein
Kidney
Umbilicus
High
Medium
Low
Umbilical arteries
Lower extremities
Internal iliac artery
Placenta

INFLUENCIA MOVIMIENTOS RESPIRATORIOS

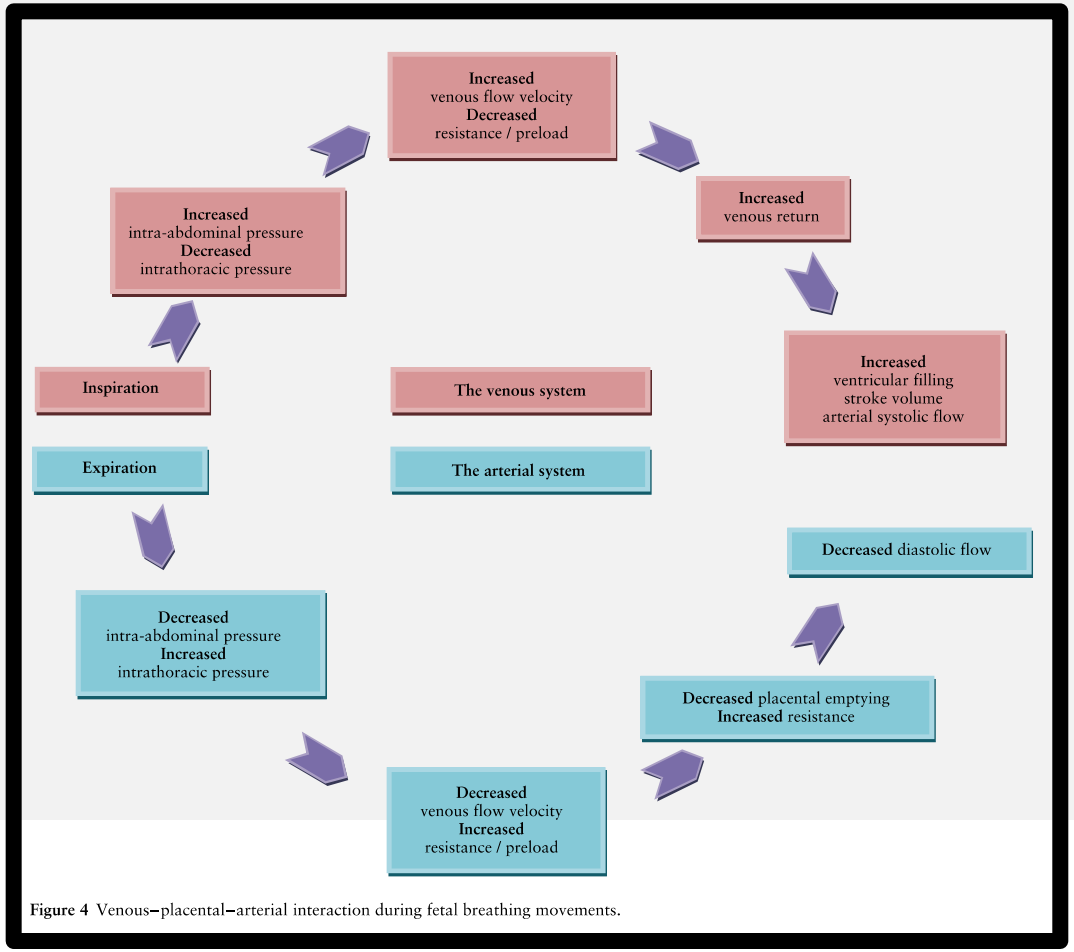


Figure 4 Venous–placental–arterial interaction during fetal breathing movements.

AGENESIA DE DUCTUS VENOSO

- FALLA DE ANASTOMOSIS CRITICA ENTRE LA VENA UMBILICAL Y EL DUCTUS VENOSO, DRENANDO POR UN VASO ABERRANTE INTRA O EXTRAHEPATICO
- SU INCIDENCIA ES RARA , CALCULANDOSE EN 6/1000 FETOS
- SE ASOCIA FRECUENTEMENTE CON ANOMALIAS CARDIACAS EXTRACARDIACAS Y CROMOSÓMICAS
- CUANDO SE ASOCIA A ANOMALIAS CROMOSOMICAS Y MALFORMACIONES TIENE UNA ALTA MORTALIDAD

AGENESIA DE DUCTUS VENOSO

INTRAHEPÁTICA

VENA UMBILICAL DRENA DIRECTO A LOS SINUSOIDES HEPÁTICOS SIN MEDIAR UN VASO .

EXTRAHEPATICA

VENA UMBILICAL DRENA A UN VASO EXTRAHEPÁTICO MEDIANTE UN CONDUCTO ABERRANTE

VCS, VCI, VIL, DIRECTO A AD

AGENESIA DE DUCTUS VENOSO

Prenatal Diagnosis of Agenesis of Ductus Venosus: A Retrospective Study of Anatomic Variants, Associated Anomalies and Impact on Postnatal Outcome

Pränatale Agenesie des Ductus venosus: retrospektive Analyse anatomischer Varianten, assoziierter Fehlbildungen und Einfluss auf das postnatale Outcome

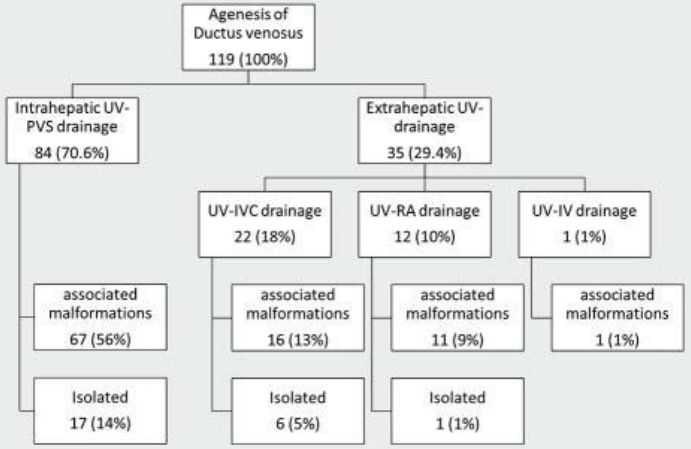
Authors
Brigitte Strizek[1, *], Aikaterini Zamprakou[1, *], Ingo Gottschalk[2], Maria Roethlisberger[2], Astrid Hellmund[1], Andreas Müller[3], Ulrich Gembruch[1], Annegret Geipel[1], Christoph Berg[2]

AGENESIA DE DUCTUS VENOSO

Purpose To assess the anatomic variants, associated anomalies and postnatal outcome of fetuses with a prenatally diagnosed agenesis of ductus venosus (ADV).

Results In 24 cases (20.2 %) ADV was an isolated finding, while 95 cases (79.8 %) had associated anomalies. We identified 84 cases (70.6 %) with intrahepatic and 35 cases (29.4 %) with extrahepatic drainage of the umbilical vein. 58.8 % of neonates were alive at follow-up. There was no statistical association between drainage site and associated anomalies or outcome. Postnatal outcome was determined by the presence and severity of associated anomalies. There was no adverse outcome in the isolated group related to ADV. Overall, there were 6 persistent portosystemic shunts, 3 of them with a spontaneous closure, and one total agenesis of the portal venous system with lethal outcome.

Conclusion Postnatal outcome in cases with ADV mainly depends on the presence of associated anomalies. In isolated cases the prognosis is generally good, but neonates with a prenatally diagnosed portosystemic shunt should be followed until its occlusion. Portal venous system agenesis is rare but should be ruled out on prenatal ultrasound.



▶ **Fig. 5** Distribution of the different umbilical venous drainage sites in 119 cases with agenesis of the ductus venosus in cases with or without associated anomalies. RA: right atrium; IV: iliac vein; IVC: inferior vena cava; UV: umbilical vein

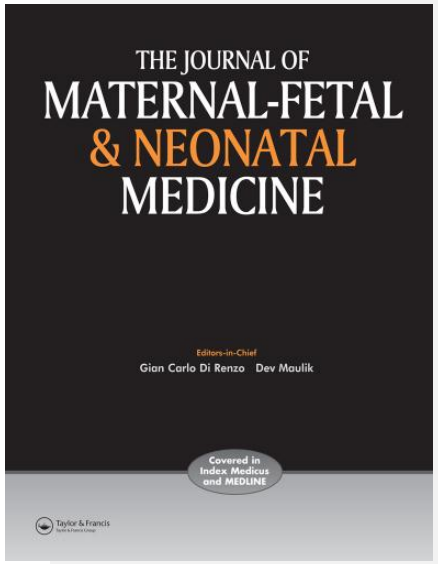
AGENESIA DE DUCTUS VENOSO

► **Table 1** Associated anomalies and outcome in 119 prenatal cases with agenesis of the ductus venosus.

associated condition		total	TOP	IUFD	NND	ICD	alive
isolated		24	0	0	1	0	23
chromosomal anomalies		24					
	trisomy 21	10	8				2
	monosomy X	5	2		1		2
	trisomy 18	2	1		1		
	other	7	4	2			1
genetic syndromes		2	1				1
	achondroplasia	1					1
	Beckwith-Wiedemann	1	1				
multiple malformations		25	11	1	4	1	8
	VACTERL	4	1		1		2
	other	21	10	1	3	1	6
cardiac anomalies only		25	2			1	22
	VSD	5				1	4
	CoA	5					5
	other	15	2				13
single extracardiac anomalies only		14	1		3		10
	diaphragmatic hernia	6			1		5
	lower urinary tract obstruction	2					2
	omphalocele	3	1				2
	hydrothorax	2			1		1
	meconium peritonitis	1			1		
cytomegalovirus infection		2	1				1
twin to twin transfusion syndrome/TRAP		3		2		1	
total		119	31	5	10	3	70

ICD: death in infancy or childhood; IUFD: intrauterine fetal death; NND: neonatal death; TOP: termination of pregnancy; TRAP: twin reversed arterial perfusion.

AGENESIA DE DUCTUS VENOSO



Original Research

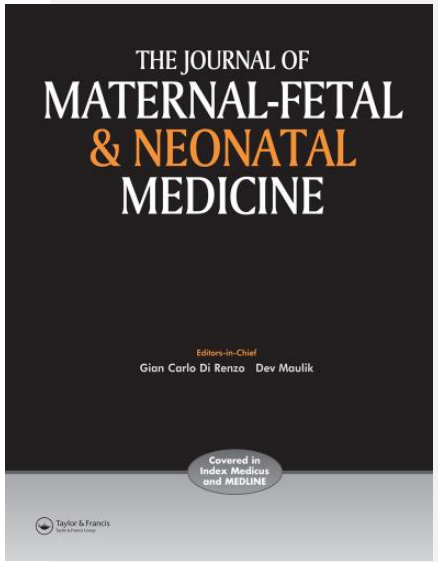
Predicting outcome in 259 fetuses with agenesis of ductus venosus – a multicenter experience and systematic review of the literature

Amirhossein Moaddab, M.D, Gabriele Tonni, MD, PhD, Gianpaolo Grisolia, MD, Maria Paola Bonasoni, MD, Edward Araujo Júnior, MD, PhD, Lilliam Cristine Rolo, MD, PhD, Federico Prefumo, MD, PhD, Sergio de la Fuente, MD, Waldo Sepulveda, MD, Nancy Ayres, MD, Rodrigo Ruano, MD PhD

Objective: To evaluate prenatal predictors of postnatal survival in fetuses with agenesis of ductus venosus (ADV).

Methods: This retrospective study reviewed our experience and the literature between 1991 and 2015. Prenatal findings were evaluated and perinatal morbidity and mortality was documented.

AGENESIA DE DUCTUS VENOSO



Results: A total of 259 cases were included in the present analysis from our centers and 49 published studies (15 patients from our retrospective cohort review and 244 from literature review). The intrahepatic and extrahepatic shunts were present in 32.0% (73/226) and 67.7% (153/226), respectively. Cardiomegaly (n=64/259, 24.7%), hydrops (n=31/259, 12.0%) and amniotic fluid abnormalities (n=22/259, 8.5%) were among the most frequent initial ultrasound findings. 147 fetuses (56.8%) had ADV without structural anomalies while 112 (43.2%) had associated anomalies (cardiac anomalies (n=66), extra-cardiac anomalies (n=19), and both cardiac and extra-cardiac anomalies (n=27). The mean gestational age at ultrasound diagnosis was 22.9±6.9 weeks while the mean gestational age at delivery was 34 ± 7.5 weeks. The overall neonatal survival was 57.1% (n=148/259). The following factors were associated with survival: advanced maternal age, earlier gestational age at diagnosis, prematurity, increased NT, pericardial effusion, associated cardiac defects (specially AVSD), chromosomal abnormalities, hydrops, hygroma, and limb anomalies.

Conclusion: Fetal hydrops, presence of associated congenital anomalies, and premature delivery are associated with poor prognosis in fetuses with ADV.

AGENESIA DE DUCTUS VENOSO

AGENESIA DE DUCTUS VENOSO : DIAGNÓSTICO PRENATAL, RESULTADO PERINATAL Y REVISION SISTEMÁTICA

REVISION RETROSPECTIVA DE TODOS LOS CASOS CON DIAGNÓSTICO PRENATAL DE ADV EN TRES CENTROS TERCIARIOS

SE COMPARA EL TIPO DE SHUNT, ASOCIACIÓN A ANEUPLOIDIA Y RESULTADO PERINATAL

SE REALIZÓ UNA REVISION SISTEMÁTICA DE 1990 A 2018 SELECCIONÁNDOSE TODAS AQUELLAS SERIES CON MAS DE 10 CASOS

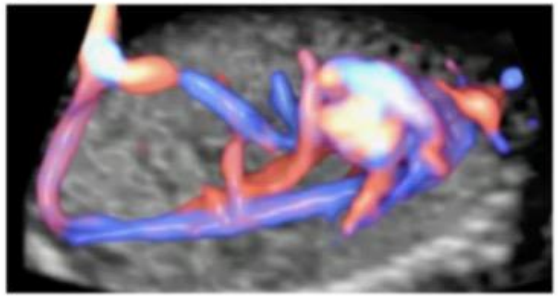
AGENESIA DE DUCTUS VENOSO

Agenesis of the Ductus venosus: Prenatal diagnosis, perinatal outcomes and Systematic review

Muñoz H, Copado DY, Aguilera S, Campanella C, Perez A, De La Fuente S, Muñoz G, Solari C, Silva MC, Parra MC, Acevedo S. **Universidad de Chile. Clinica Las Condes. Instituto Nacional de Perinatologia Isidro Espinosa De los Reyes, Mexico City, Mexico.**

Introduction: The Ductus venosus connects the umbilical vein with the left portal system and the left hepatic vein, allowing proper oxygenation of the fetal organs. First case of Agenesis of ductus venosus (ADV) was described in 1991. Incidence is 1:2500, in 11 to 14 weeks examination.

Objective: The aim of this study is to describe the prenatal diagnosis and perinatal outcomes in fetuses with agenesis of the ductus venosus (ADV) including shunting type, morphological and chromosome-associated pathology, and perinatal outcome. And perform systematic review of literature.



AGENESIA DE DUCTUS VENOSO

34 CASOS DE ADV FUERON DIAGNOSTICADOS DURANTE ESTE PERIODO , LA EDAD MATERNA PROMEDIO FUE DE 27 AÑOS (16-40) Y LA EDAD GESTACIONAL PROMEDIO AL DIAGNÓSTICO FUE DE 21 SEMANAS(11-37 SEM)

SE REVISARON 283 CASOS DE ADV RECOLECTADOS DE 10 PUBLICACIONES

AGENESIA DE DUCTUS VENOSO

Associated anomalies and outcome in 34 prenatal cases with genesis of the ductus venosus

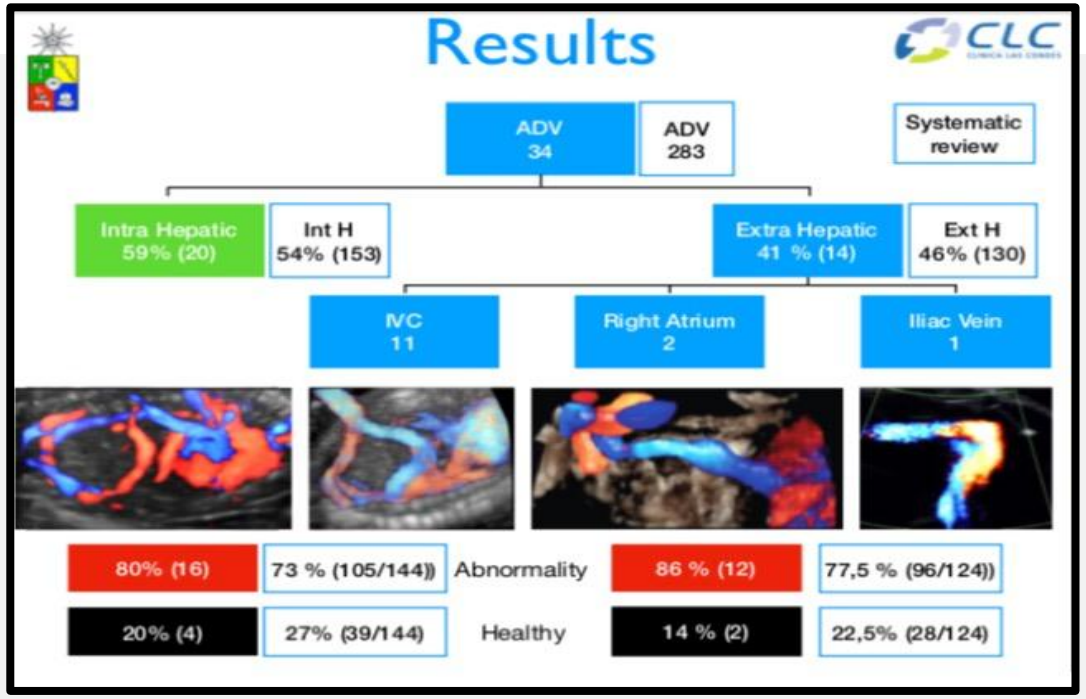
Associated condition		Total	Systematic review	Alive healthy	Associate anomaly alive	lost/ ongoing	Perinatal Mortality
Isolated		17,6 % (6)	23,7 % (67/283)	17,6 % (6/34)	-	-	-
Chromosomal or genetic anomalies		32,4 % (11)	18,7 % (53(283)	-	3	2	6
	Monosomy X	9					
Multiple or extracardiac anomalies only		29,4% (10)	34,6% (98/283)	—	2	1	7
	Diaphragmatic hernia	2					
Cardiac anormalies only		20,6% (7)	23% (65/283)		6	1	-
	VSD	1					
Total		34		17,6 % (6/34)	32,4 % (11/34)	11,8 % (4/34)	38,2 % (13/24)

AGENESIA DE DUCTUS VENOSO

ADV 34 CASOS

	11-14 47% (16)	20 SEM 53% (18)	REVISION 283
ANOMALIA	31% (5)	66% (12)	50 % 17	57,6 % 163
ANEUPLOIDIA	50% (8)	17% (3)	32,4% (11)	18,7 % (53)
SANOS	20% (3)	17% (3)	17,6% (6)	23,7 % (67)

AGENESIA DE DUCTUS VENOSO



AGENESIA DE DUCTUS VENOSO

LA AGENESIA DE DUCTUS VENOSO ES UNA CONDICION POCO FRECUENTE

TIENE UNA ALTA ASOCIACIÓN A MALFORMACIONES Y ANEUPLOIDIAS . ALTA MORTALIDAD

CUANDO SE ENCUENTRA DE MANERA AISLADA EN GENERAL ES DE BUEN PRONÓSTICO

DEBE SER BUSCADA DE MANERA ACTIVA EN LA ECOGRAFÍA 11-14 SEMANAS