PENTALOGIA DE CANTRELL

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Sinónimos

- Síndrome de Cantrell
- Síndrome de Cantrell-Haller- Ravitch
- Hernia Peritoneo-Pericardio-Diafragmática
- Síndrome Toracoabdominal
- Ectopia Cordis Toracoabdominal

Generalidades

- Descrita por J.R Cantrell en 1958.
- Poco frecuente.
- Casos publicados: 100
- Incidencia: 5.5 por 1 millón RNV.
- Predominio de sexo masculino 2:1

Definición

- Conjunto de malformaciones que se caracterizan por:

1. Defectos del tercio inferior del esternón
2. Defectos de la pared anterior en la línea media epigástrica
3. Defectos del diafragma anterior
4. Defecto del pericardio diafragmático
5. Defectos cardiacos

Etiología

- Desconocida
- Aneuploidias
- Herencia dominante ligada al cromosoma Xq 25-26
- ⇩ Reducción de la expresión de BMP2
- Infección viral??????
- Teratógenos: quinidina, warfarina, talidomina
- Exposición sustancias toxicas: amino-propionitrilo
- Deficiencia de vitamina A

Patogénesis

Alteración del desarrollo mesodérmico

Defectos Pericardico

Defectos Cardiacos

Defectos Diafragmáticos

Migración ventral inadecuada de estructuras del primordio

Defectos del esternón

onfalocele

Clasificación

- Toyama en 1972, describió 3 variantes:
- Clase 1 (Diagnostico Certero): 5 defectos presentes
- Clase 2 (Diagnostico Probable): 4 defectos presente (incluye ectopia cordis y defectos de la pared abdominal)
- Clase 3 (Diagnostico Incompleto): combinaciones variables de los defectos (incluye anomalías del esternón)

DIAGNOSTICO

- PRENATAL

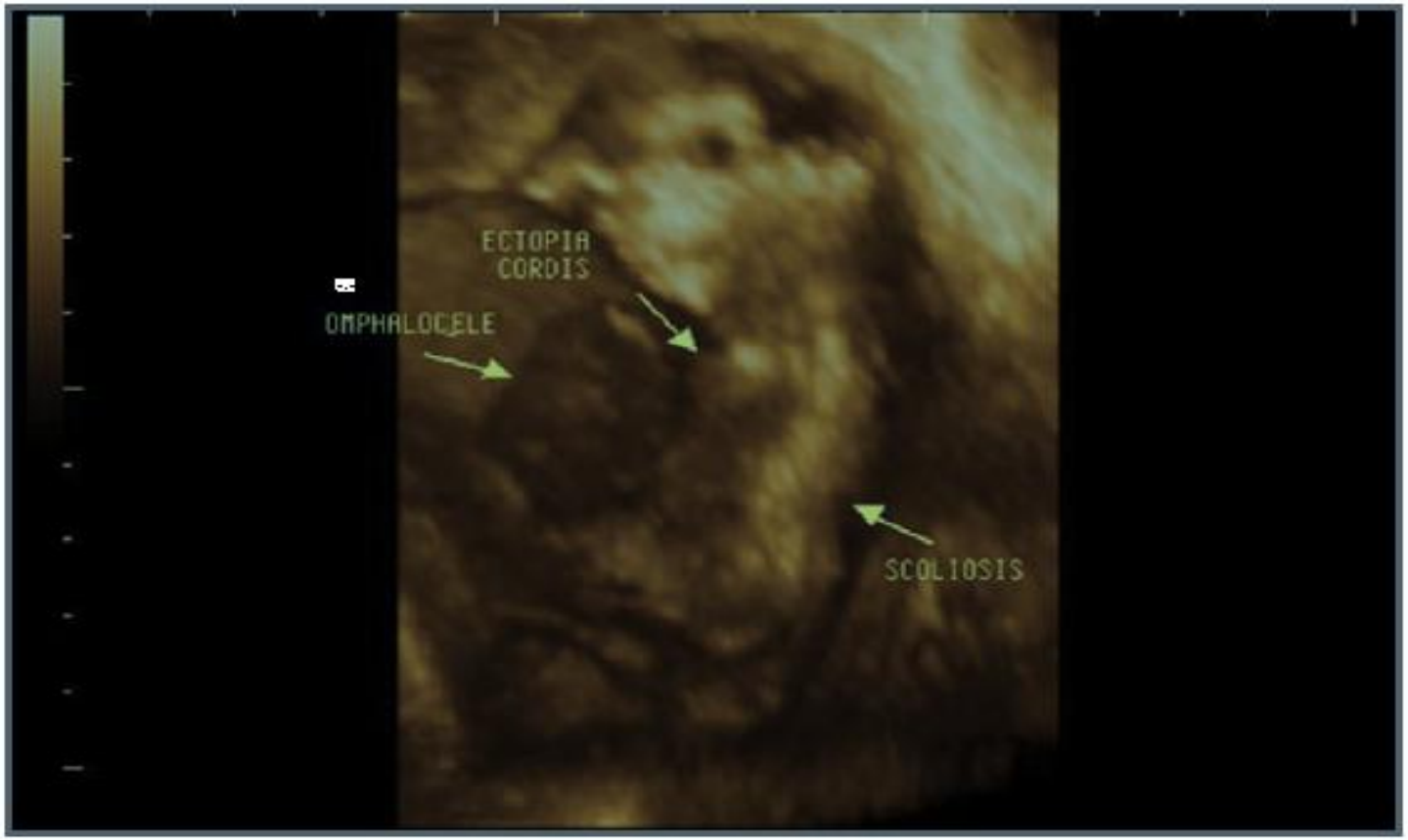
US

- ECO 11-14: Onfalocele, quiste alantoideo, ectopia cordis, TN aumentada, Doppler DU onda A(-)
- ECO 18-24: agenesia parcial o total del esternón, Ectopia cordis, defectos cardiacos(CIV, CIA, CAV, Diverticulos ventriculares, TGA, ventriculo unico, drenaje pulmonar anomalo, tronco pulmonar hipoplasico, T.Fallot), Hernia diafragmatica, onfalocele, hidrops, alteraciones del doppler.

RMF

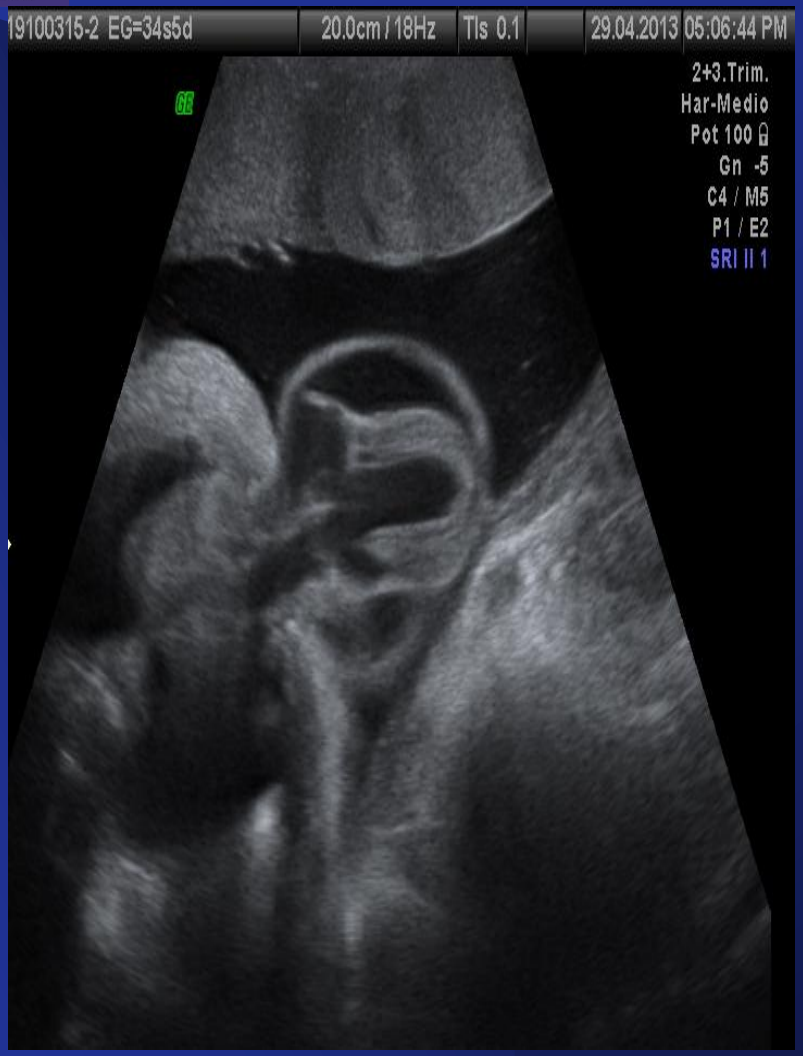
- Confirmación del diagnostica.
- Obtención de imágenes de estructuras anatómicas fetales.

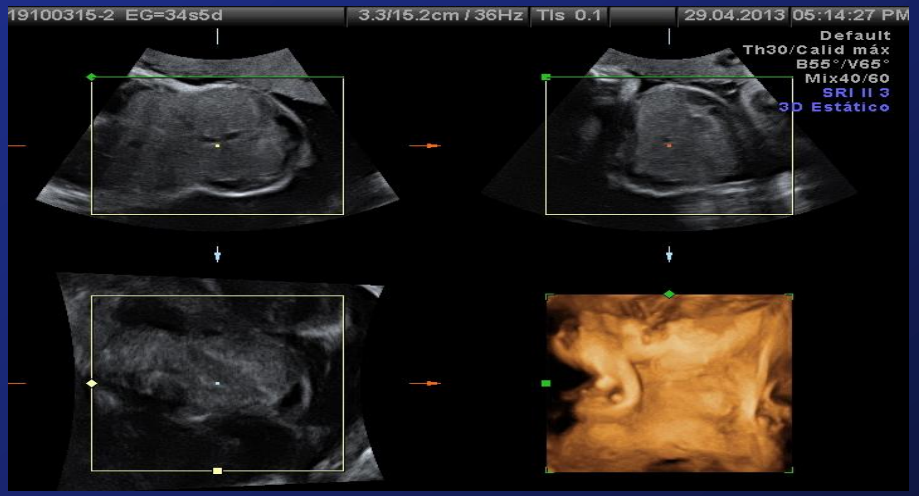
ECO 3D

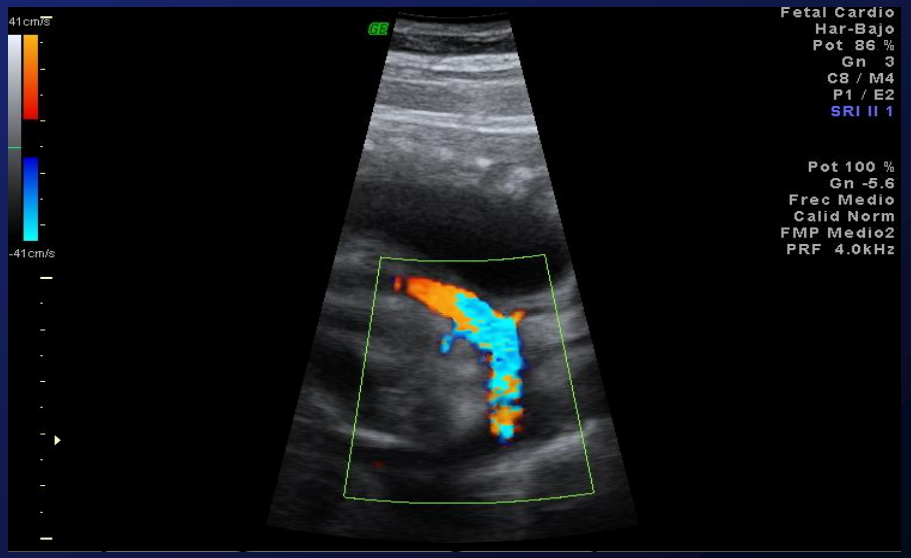


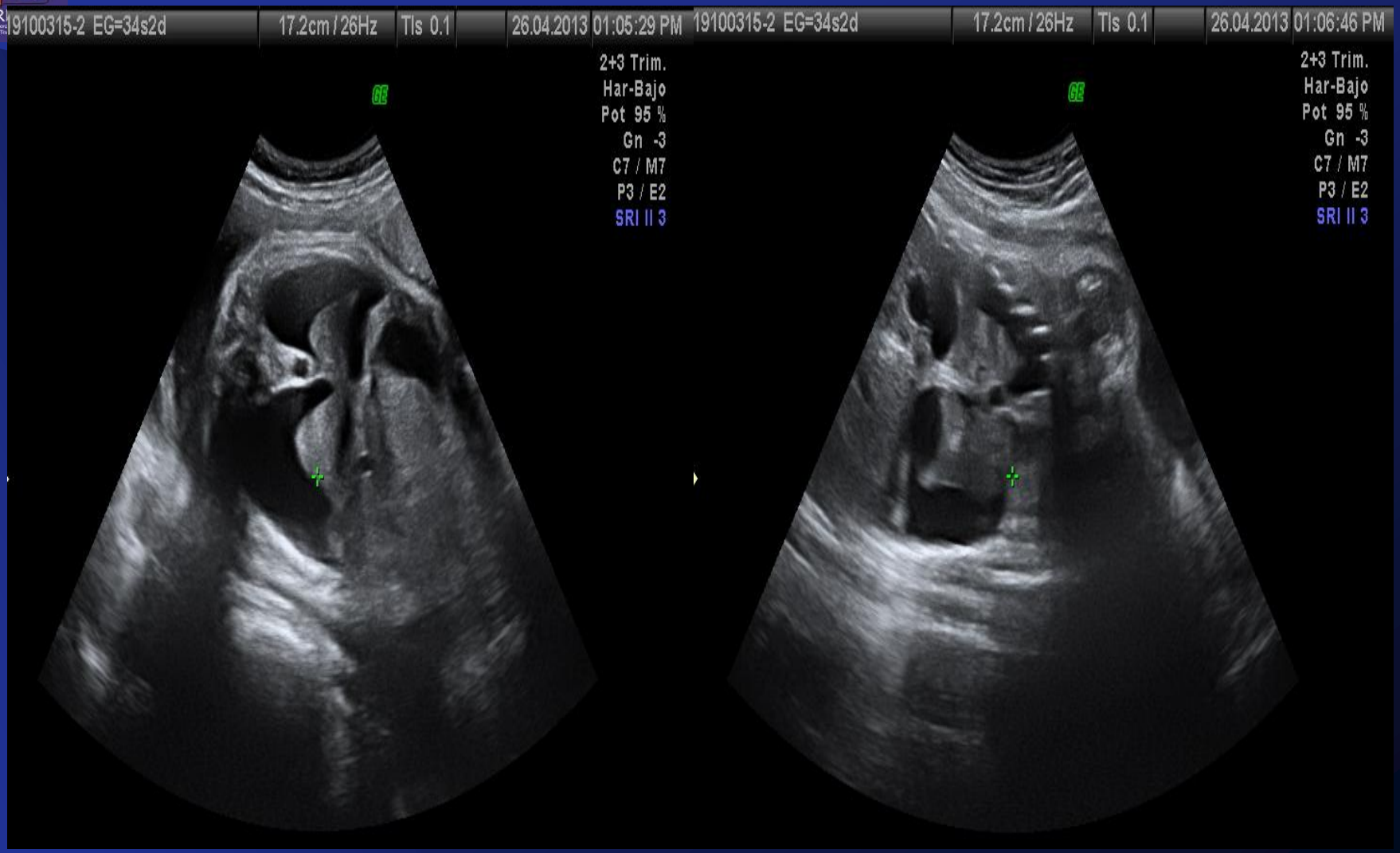
The ectopia cordis, omphalocele, and marked spinal curvature were clearly observed by 3D US with surface mode.

Ultrasonografía





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2+3 Trim.
Har-Bajo
Pot 95 %
Gn -3
C7 / M7
P3 / E2
SRI II 3
GE
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17.2cm / 26Hz
Tls 0.1
26.04.2013 01:06:46 PM
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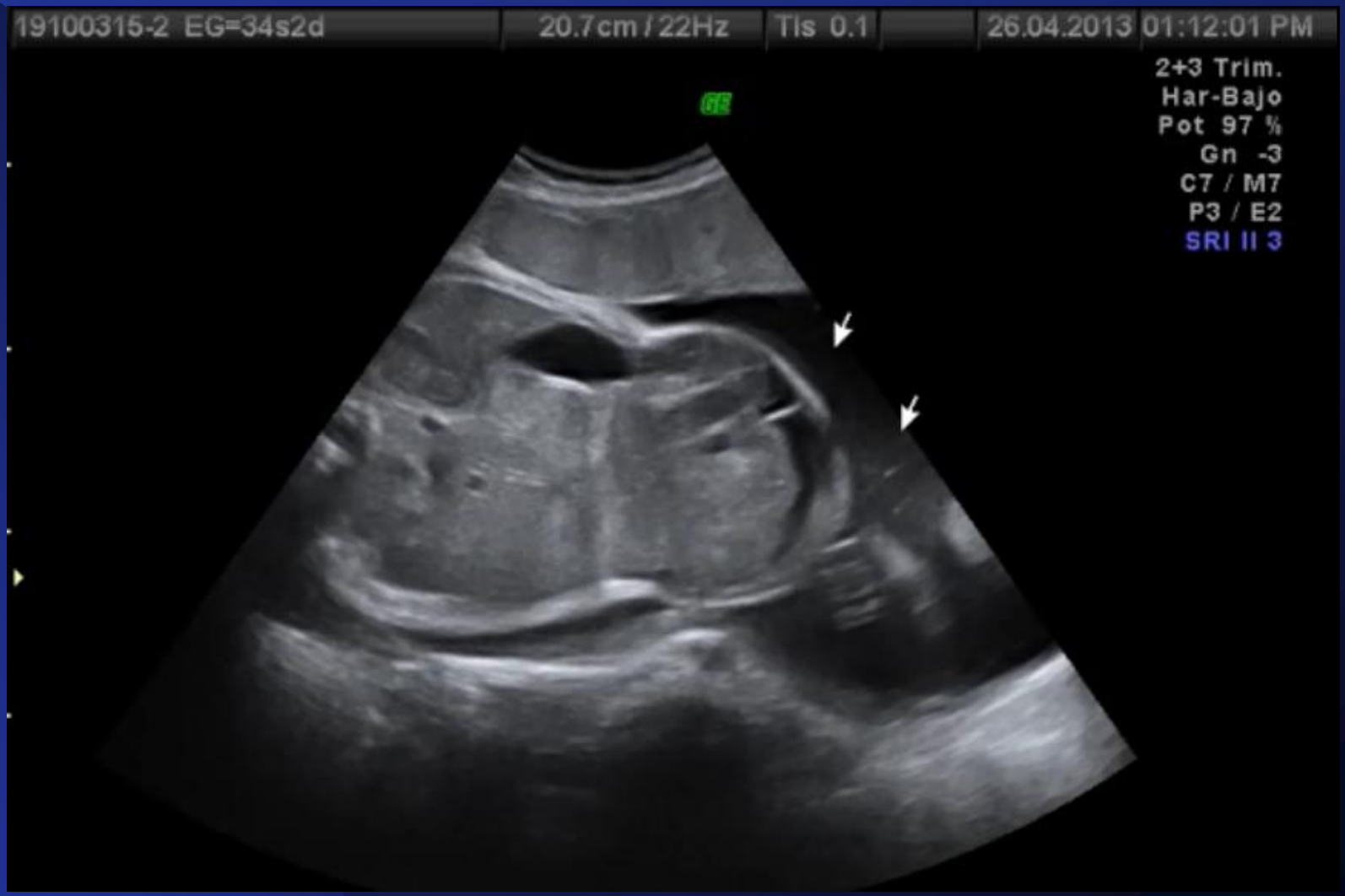
Har-Bajo
Pot 95 %
Gn -3
C7 / M7
P3 / E2
SRI II 3
GE

US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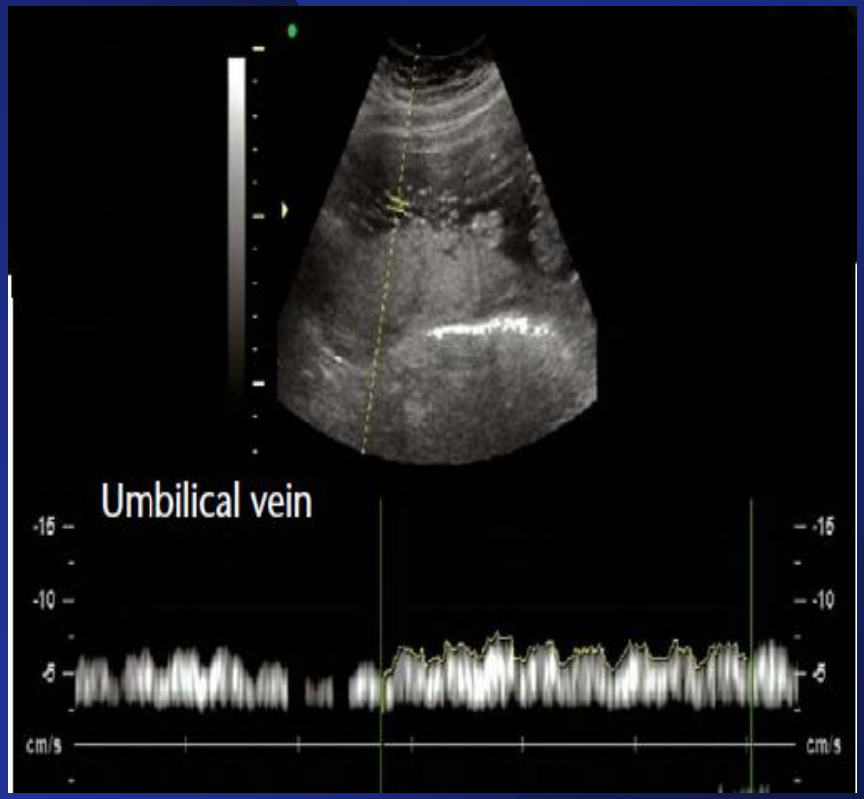
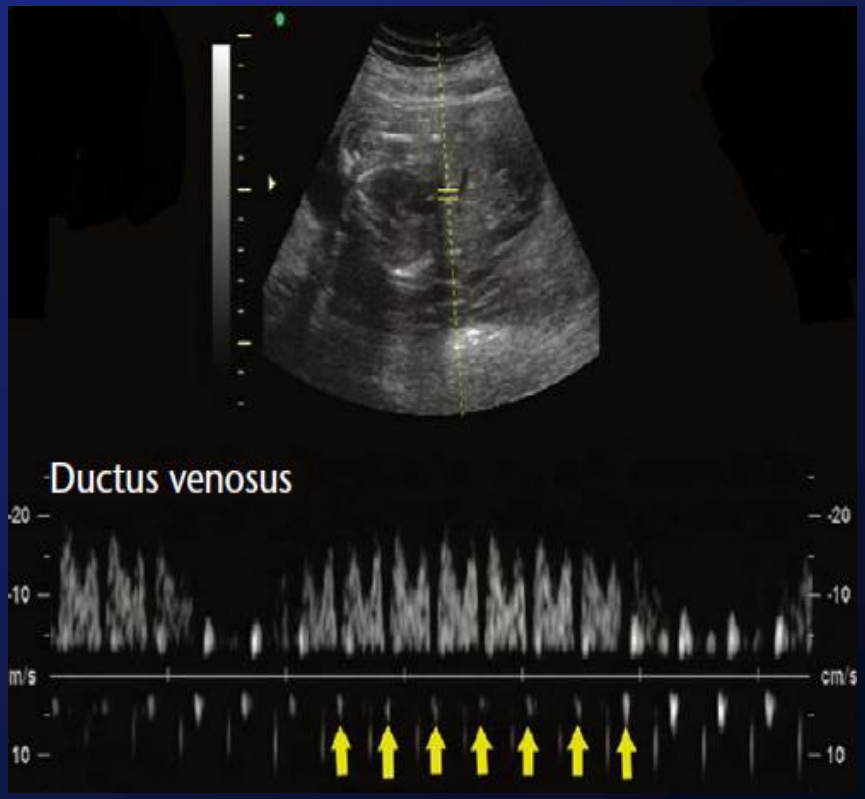
Har-Bajo
Pot 97 %
Gn -3
C7 / M7
P3 / E2
SRI II 3

Alteraciones del Doppler

Vena umbilical

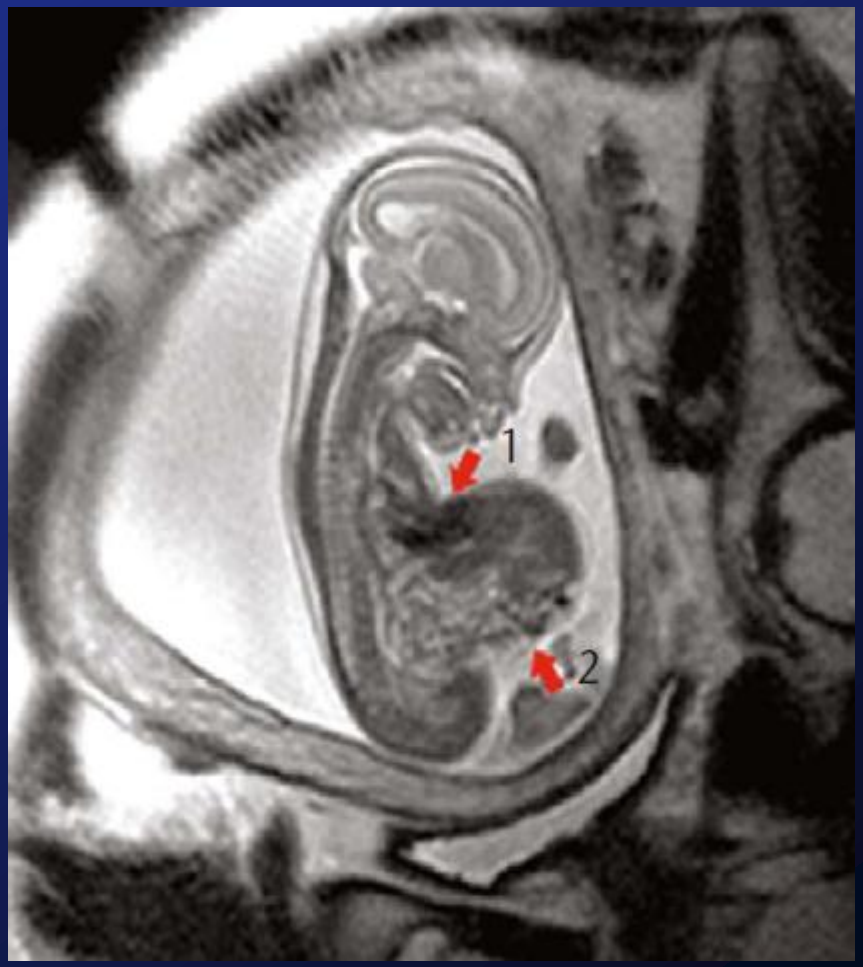


Ductus venoso



RMF

- 1. Defecto bajo del esternón, pared abdominal, ectopia cordis
- 2. onfalocele contenido hepático e intestinal



Malformaciones Asociadas

- SNC: encefalocele, siringomielia, espina bífida, exencefalia, ventriculomegalia, craneosinostosis
- Faciales: dirrinia, labio leporino, fisura palatina, orejas de implantación baja
- Cuello: higroma quístico
- TGI: poliesplenia, divertículo de Meckel, atresia colonica o ano.
- GU: riñón en herradura, cambios multiquísticos renales, megavejiga.
- Esqueléticas: escoliosis severa, pie bot, clinodactilia, sirenomelia
- Polihidroamnios y arteria umbilical única.

Diagnósticos Diferenciales

- Síndrome de Banda amniótica
- Onfalocele aislado
- Ectopia Cordis
- Síndrome del Cordon corto
- Síndrome de Beckwith-Wiedemann

Vía del parto

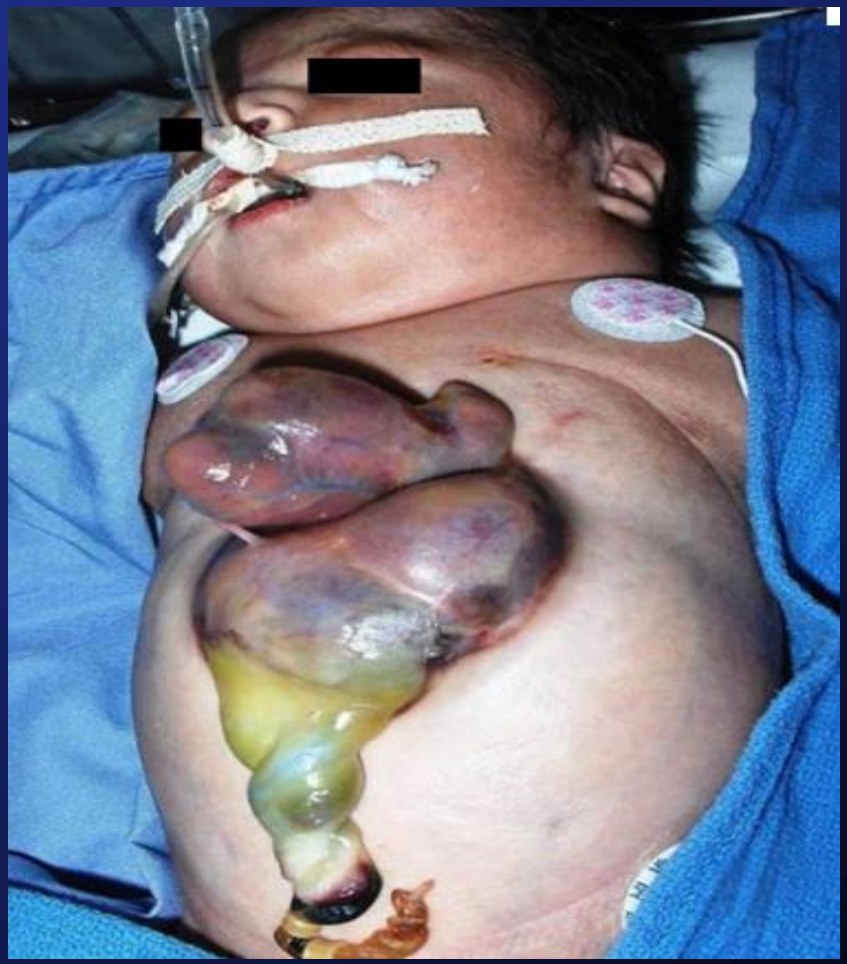
- El parto por vaginal o cesárea.
- Cesárea disminuye el riesgo de infección y la probabilidad de ruptura visceral.

Pronostico

- Cariotipo
- Parto prematuro y restricción de crecimiento fetal 30%
- Mortalidad al nacimiento 50%, con ectopia cordis cervical 100%
- Supervivencia del 20% sin defectos cardiacos y variantes incompletas.
- Supervivencia del 8% con P.cantrell completa.

Manejo

- Proteger las órganos expuestos
- Manejo ventilatorio, hemodinámico, electrolítico y antibióticos de amplio espectro
- Manejo multidisciplinario
- Tratamiento quirúrgicos correctivos por etapas (defectos cardiacos, torácicos, abdominales)
- Resultados controversiales



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Surgical treatment of three cases of Cantrell's syndrome

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Abstract

This ten-year retrospective study was designed to examine the morbidity and mortality of three cases of Cantrell's syndrome between 1998 and 2008. The three patients showed different degrees of Cantrell's pentalogy including abdominal ectopia cordis, thoracic-abdominal ectopia cordis and left ventricular diverticulum. Of the three, the 5-month-old boy suffering from complicated congenital heart disease with abdominal ectopia cordis received a successful single stage repair and reconstruction of the abdominal wall. The 33-week-old premature girl with thoracic-abdominal ectopia cordis underwent two stage correction of tetraology of Fallot. The 4-year-old girl underwent ectomy of left ventricular diverticulum and thoracoabdominal wall repair. Twenty-four to thirty-five months follow-up were satisfactory. We hold that two-stage repair are technically feasible for Cantrell's syndrome, especially for those with complex congenital heart diseases. Post-operative ventilatory support and multiple post-operative care should be prolonged. Malnutrition, infection and arrhythmia are central problems in medical care and surgery should be considered if there was progressive heart failure or hemodynamic instability.

Resultados

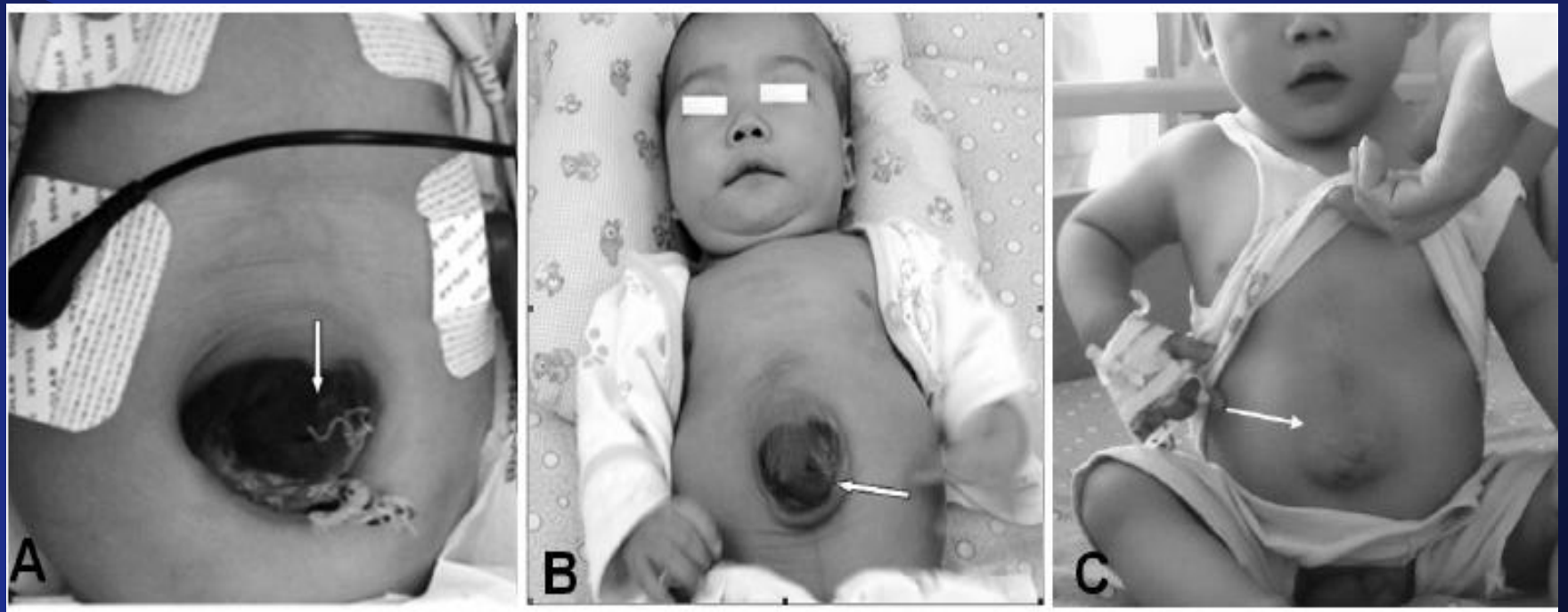

Fig 1. A. The abdominal ectopia cordis covered with a tiny, ragged and necrotic skin. B. The malnutrition and ragged skin of ectopia cordis turned better after considerate treatment. C. The ectopia cordis submerged into abdominal cavity and the patient was asymptomatic without restriction in activity at the 24-month follow-up.

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