

Centro de Referencia Perinatal Oriente

Facultad de Medicina, Universidad de Chile



Manejo del embarazo gemelar monocorial monoamniótico

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Becada Obstetricia y Ginecología HLTB, U. Chile.

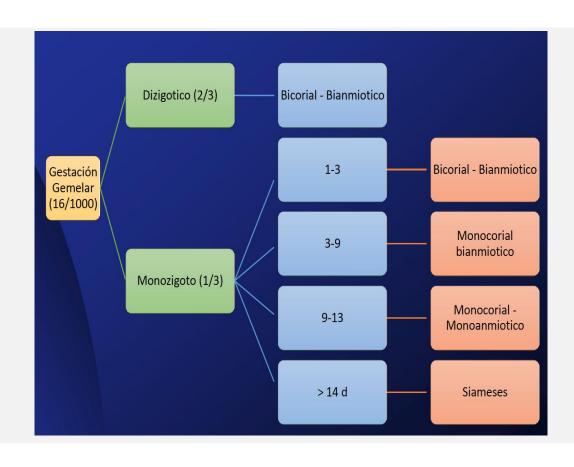
26 de Junio de 2018



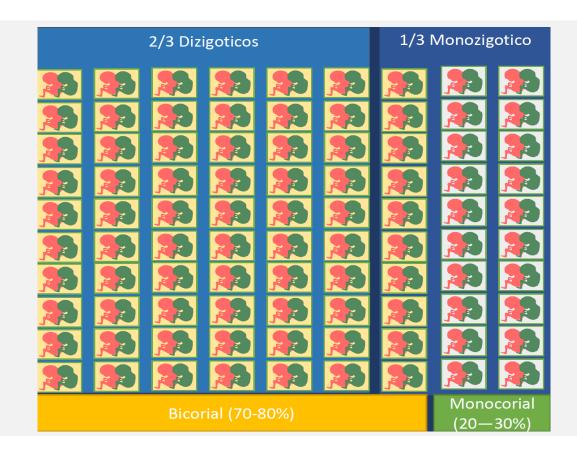
- Los embarazos gemelares representan un 1,4% de todos los embarazos.
- 20% consisten en gemelos monocoriales y un 5% de éstos son monoamnióticos.
- Existe un incremento del embarazo múltiple en los últimos 30 años.
 - Terapia de reproducción asistida y edad materna avanzada.

Newman. Multiple Gestations: Timing of Indicated Late Preterm and Early-Term Births in Uncomplicated Dichorionic, Monochorionic, and Monoamniotic Twins. Semin Perinatol 35:277-285 © 2011.











Riesgos perinatales significativos:

	ВС	MC
Aborto (11-23 sem)	2%	10%
Muerte perinatal (>23 sem)	2%	4%
RCF (≥1)	20%	30%
Parto prematuro(<32 sem)	5%	10%
Defectos mayores	1%	4%

Newman. Multiple Gestations: Timing of Indicated Late Preterm and Early-Term Births in Uncomplicated Dichorionic, Monochorionic, and Monoamniotic Twins. Semin Perinatol 35:277-285 © 2011.



- Los gemelos monoamnióticos ocurren en un 1% de todos los embarazos gemelares monocigóticos.
- La incidencia varia 1 en 1650 a 1 en 90.000 nacimientos (1:10.000).
- 9 veces más anomalías congénitas que un embarazo único (18-28%). 2-3% STFF.

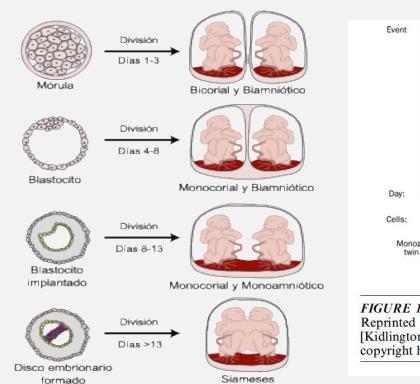
Post. Managing Monoamniotic Twin Pregnancies. CLINICAL OBSTETRICS AND GYNECOLOGY Volume 58, Number 3, 643–653 2015.



 Baja prevalencia, por lo que la mayoría de la evidencia proviene de series pequeñas y reportes de casos.

Post. Managing Monoamniotic Twin Pregnancies. CLINICAL OBSTETRICS AND GYNECOLOGY Volume 58, Number 3, 643–653 2015.





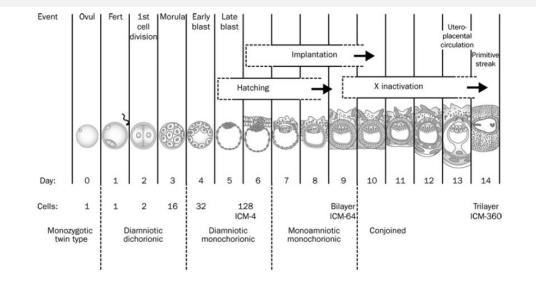


FIGURE 1. Schematic drawing of human embryonic development with timing of twinning. Reprinted from Hall.² Copyright 2003, with permission from Elsevier. Copyright [Elsevier], [Kidlington, Oxford, UK]. All permission requests for this image should be made to the copyright holder.

Newman. Multiple Gestations: Timing of Indicated Late Preterm and Early-Term Births in Uncomplicated Dichorionic, Monochorionic, and Monoamniotic Twins. Semin Perinatol 35:277-285 © 2011.



- 50% reportes antiguos: Parto prematuro, RCIU, anomalías congénitas, STFF y en un 50% cordón umbilical enredado.
- 5-20% actualmente: diagnóstico temprano, uso de corticoides antenatales, incremento de vigilancia fetal, mejora del cuidado neonatal e interrupción electiva temprana.

Newman. Multiple Gestations: Timing of Indicated Late Preterm and Early-Term Births in Uncomplicated Dichorionic, Monochorionic, and Monoamniotic Twins. Semin Perinatol 35:277-285 © 2011.



- En un review entre 1990 y 2002 se reporta 23% de mortalidad perinatal.
- Un 50% de las muertes intrauterinas por causa de la oclusión de cordón umbilical.
- Se estima que un 25% de éstos embarazos sufren al menos una pérdida después de las 20 semanas.



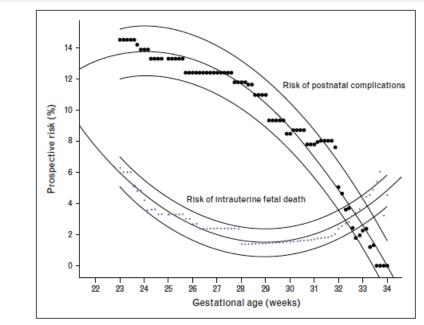
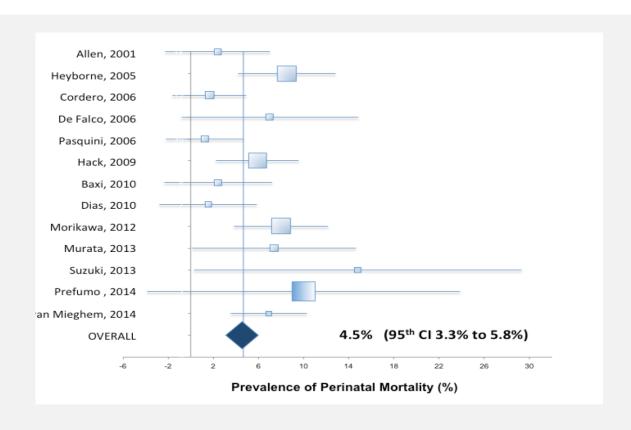


FIGURE 1. Regression line (and 95% CI) of the prospective risk of intrauterine fetal death and postnatal complications between 23 and 34 weeks of gestation. The left and right angles of the polygon formed by the intersection of the 95% CIs of the prospective risk of intrauterine fetal death and the risk of a postnatal complication determine the 95% CI of the 'optimal time of delivery.' CI, confidence interval. Reproduced from [8*].

Ishii. Prenatal diagnosis and management of monoamniotic twins. Copyright © 2015 Wolters Kluwer Health.





Prefumo. The natural history of monoamniotic twin pregnancies: a case series and systematic review of the literature. 2015



- Se cree una de las principales causas de morbimortalidad de los gemelares monoamnióticos.
- 88% de prevalencia en gemelares monoamnióticos y en un 65% de muertes fetales¹.
- La mayoría de los casos ocurre en periodo previable.

1. Murata M, Ishii K, Kamitomo M, et al. Perinatal outcome and clinical features of monochorionic monoamniotic twin gestation. J Obstet Gynaecol Res 2013; 39:922–925.



Ultrasound Obstet Gynecol 2013; 41: 131–135 Published online in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/uog.12345



Impact of cord entanglement on perinatal outcome of monoamniotic twins: a systematic review of the literature

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• 9 estudios, 114 gemelares monoamnioticos (228 fetos) con cordón umbilical enlazado un 88,6% sobrevivieron. De los 26 muertos (11,4%), solo 2 fueron por un accidente de cordón.

Cordón umbilical "enlazado"



	Co entang		No cord entanglement			Odds ratio	Odds ratio	
Study or Subgroup	Events	Total	Events	Total	Weight	M-H, fixed, 95% CI	M-H, fixed, 95% CI	
Cordero ²	0	30	4	42	40.9%	0.14 (0.01, 2.71)		
DeFalco ¹¹	1	22	2	18	23.2%	0.38 (0.03, 4.58)		
Sau ¹³	4	10	2	4	18.9%	0.67 (0.06, 6.87)		
Suzuki ¹⁴	3	20	1	2	17.0%	0.18 (0.01, 3.66)		
Total (95% CI)		82		66	100.0%	0.30 (0.08, 1.10)		
Total events	8		9					
Heterogeneity: $Chi^2 = 0.85$, $df = 3$ (P = 0.84); $I^2 = 0\%$						0.01	0.1 1 10	100
Test for overall effect	ct: Z = 1.8	81, (P =	0.07)			0.01	Favors cord Favors no cord entanglement entanglement	100

Figure 2 Forest plot of odds ratio for overall mortality according to presence or absence of cord entanglement in monoamniotic twins. Numbers refer to number of fetuses. M–H, Mantel–Haenszel test.

			No cord entanglement Odds ratio				C	o			
Study or Subgroup	Events	Total	Events	Total	Weight	M–H, fixed, 95% CI	[M-H,	fixed, 9	5% CI	
Cordero ²	2	30	11	42	48.0%	0.20 (0.04, 0.99)					
DeFalco ¹¹	13	22	14	18	35.3%	0.41 (0.10, 1.67)					
Sau ¹³	4	10	2	4	9.6%	0.67 (0.06, 6.87)			-		
Suzuki ¹⁴	6	20	1	2	7.1%	0.43 (0.02, 8.04)					
Total (95% CI)		82		66	100.0%	0.34 (0.14, 0.82)			_		
Total events	25		28				1	ı		1	1
Heterogeneity: Chi ²	$= 0.84, \alpha$	df = 3 (F	0 = 0.84;	$I^2 = 0\%$			0.01	0.1	1	10	100
Test for overall effect	ct: $Z=2$.	39, (P =	0.02)					Favors cord entanglement		Favors no cord entanglement	

Figure 3 Forest plot of odds ratio for neonatal morbidity according to presence or absence of cord entanglement in monoamniotic twins. Numbers refer to number of fetuses. M–H, Mantel–Haenszel test.



- Detección de un 98% por ultrasonido, edad gestacional al diagnóstico entre las 11 y 30 semanas.
- Morbilidad 21,7%.
- No existen diferencias significativas en las tasas de mortalidad de los gemelos monoamnióticos con o sin cordón umbilical enlazado. La morbimortalidad depende principalmente del STFF, la prematuridad y anomalías congénitas.



Ultrasound Obstet Gynecol 2010; 35: 201–204
Published online 12 January 2010 in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/uog.7501

Cord entanglement and perinatal outcome in monoamniotic twin pregnancies

T. DIAS, S. MAHSUD-DORNAN, A. BHIDE, A. T. PAPAGEORGHIOU and B. THILAGANATHAN

Fetal Medicine Unit, Academic Department of Obstetrics and Gynaecology, St George's Hospital Medical School, London, UK



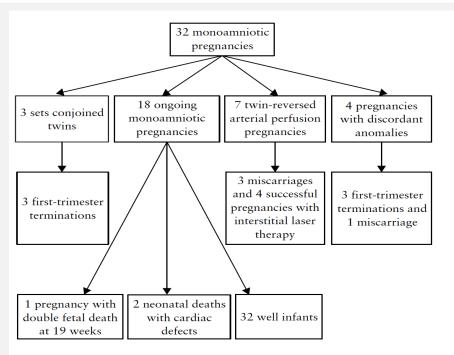


Figure 1 Flow chart showing the main prenatal diagnoses, chronology and outcome of the 32 monoamniotic pregnancies in the study.

De los 18 un 100% tenía cordón umbilical enredado diagnosticado por modo B y doppler color a las 11-16 semanas.

Mortalidad perinatal: 11,1% después de las 16 semanas y 5,9% de las 20 semanas.



Table 1 Review of the published literature since 2000 containing more than five cases and indicating prevalence of cord entanglement and perinatal outcome

Reference	Number of cases	Cord entanglement (n (%))	Timing of diagnosis of cord entanglement	IUD (n)	NND (n)	Survivors (n)	Perinatal surviva rate (%)
Sau <i>et al.</i> 2003 ⁸	7	4 (57)	Ultrasound scan	5	0	9	64
Ezra <i>et al</i> . 2005 ⁶	30	26 (87)	Ultrasound scan and at delivery	24	1	35	58
Cordero et al. 2006 ⁵	36	15 (42)	At delivery	1	5	66	92
Pasquini et al. 2006 ¹³	20	19 (95)	Ultrasound scan	0	0	40	100
Hack et al. 20099	98	Not reported	_	34	12	150	77
Current study Total	209	18 (100) 82/111 (74)	Oltrasound scan	Ž	Ž	332/418	89 79

IUD, intrauterine death; NND, neonatal death.

Diagnóstico



- El mapeo de flujo de color y la velocimetría Doppler están asociados con un valor predictivo positivo del 89% para el diagnóstico de cordón umbilical enredado.
- Se puede diagnosticar desde las 10 semanas.
- Distintas anomalías se han descrito en el doppler de AU: Aumento IR, FDA y notch.

Diagnóstico



 La presencia de notch es un fuerte predictor de anormalidad de cordón.



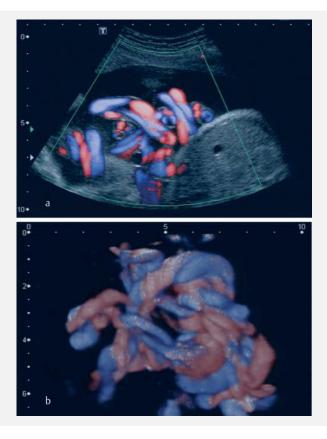


Hugon. Notching of the umbilical artery waveform associated with cord entanglement in a monoamniotic twin pregnancy. J Matern Fetal Neonatal Med, 2013; 26(15): 1559–1561

Diagnóstico



• Es de utilidad la representación 3D.

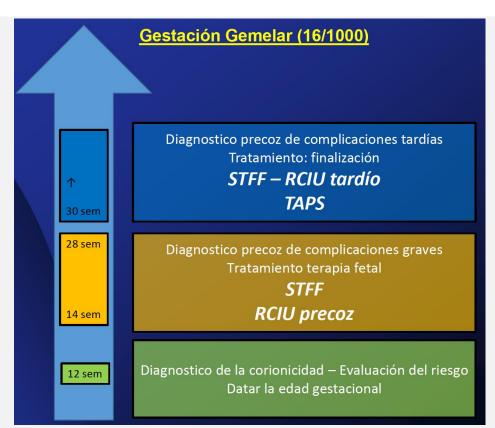


Kuwata. 3D color Doppler of monoamniotic twin cord entanglement. Arch Gynecol Obstet (2010) 281:973–974



Evaluación
 ecográfica c/1-2
 semanas.

TABLA V.										
	14	16	18	20	22	24	26	28	32	34
Biometrías + Doppler AU										
LA + vejigas + circunferencia abdominal										
Longitud cervical										





Original Research

Prenatal Management of Monoamniotic Twin Pregnancies

Tim Van Mieghem, MD, PhD, Roel De Heus, MD, PhD, Liesbeth Lewi, MD, PhD, Philipp Klaritsch, MD, Martina Kollmann, MD, David Baud, MD, PhD, Yvan Vial, MD, Prakesh S. Shah, MD, Angela C. Ranzini, MD, Lauren Mason, MD, Luigi Raio, MD, Regine Lachat, MD, Jon Barrett, MD, MBBCh, Vesal Khorsand, MD, Rory Windrim, MB, and Greg Ryan, MB

Estudio de cohorte multicéntrico. 193 gemelares monoamnióticos. 23% anomalías congénitas.

- Grupo 1: Ambulatorio desde 28 semanas.
- **Grupo 2:** Hospitalización electiva antes de las 29 semanas.
- Grupo 3: Hospitalización antes de las 29 semanas por complicaciones obstétricas.

ISSN: 0029-7844/14



- Ambulatorio:
 US + RBNE
 semanal
- Hospitalizado:
 US c/2 días +
 RBNE 3 veces
 al día.
- Corticoides entre 26-28 semanas.

Table 5. Comparison of Pregnancy Outcomes of Pregnancies With Two Fetuses Alive at 26 Weeks of Gestation by Management Setting (n=144)

Outcome	Group 1, Primary Outpatient (n=53)	Group 2, Elective Inpatient (n=71)	Group 3, Complicated (n=20)	Р
Total no. admitted	30 (56.6)	71 (100)	20 (100)	
Gestational age at admission (wk)	31.2 ± 1.8	27.5±1.1	26.6±1.6	<.01*
Gestational age when starting steroids (wk)	28.9 ± 2.0	27.6 ± 1.6	26.9 ± 1.5	<.01 [†]
Surveillance				
Cardiotocograms/wk	4.2 ± 5.5	16.6 ± 4.8	16.1 ± 6.5	<.01 [†]
Ultrasonograms/wk	1.5 ± 1.0	2.2 ± 0.6	2.3 ± 1.8	<.01 [†]
Reason for delivery				
Elective	25 (47.2)	45 (63.4)	4 (20)	<.01 [‡]
Nonreassuring fetal status	12 (22.6)	15 (21.1)	7 (35)	.45
Indicated delivery	13 (24.5)	10 (14.1)	8 (40)	.04
Termination of pregnancy for IUFD	3 (5.7)	1 (1.4)	1 (5)	.34
vaginal delivery (including	3 (5./)	1 (1.4)	∠ (10)	.15
termination of pregnancy)				
Any IUFD	3 (5.7)	1 (1.4)	1 (5)	.34
Single IUFD	1 (1.9)	0 (0)	1 (5)	.12
Double IUFD	2 (3.8)	1 (1.4)	O (O)	.73
Gestational age delivery (wk, live births)	33.0±1.8	32.2±1.2	30.7±1.9	<.01*

IUFD, intrauterine fetal death.

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ISSN: 0029-7844/14



- Ambulatorio: US + RBNE semanal
- Hospitalizado:
 US c/2 días +
 RBNE 3 veces
 al día.
- Corticoides entre 26-28 semanas.

Table 6. Neonatal Outcomes of Liveborn Neonates From Pregnancies in Which Two Fetuses Were Alive at 26 Weeks of Gestation (n=280)

Outcome	Group 1, Primary Outpatient	Group 2, Elective Inpatient	Group 3, Complicated	P
Total cohort	101	140	39	
Female sex	75 (72.8)	96 (68.1)	25 (64.1)	.36
Birth weight (g)	1.827 ± 407	1.776±291	1,585±384	<.01
Birth weight percentile	38±31	46±28	52±34	.25
Birth weight less than the 3rd percentile	16 (15.8)	2 (1.4)	2 (5.1)	<.01*
Apgar score at 5 min less than 7 (n, %)	16 (16)	7 (1.6)	0 (0)	<.01*
Nonanomalous neonates (n)	96	135	35	
Apy nonrespiratory neonatal	9 (9 4)	13 (9.6)	12 (34 3)	< 01 [†]
complication				
Neonatal death	3 (3.1)	1 (0.7)	1 (2.9)	.33
Sepsis in survivors	3 (3.2)	13 (9.6)	9 (26.5)	<.01 [†]
Intraventricular hemorrhage in survivors	1 (1.0)	0 (0)	3 (8.8)	<.01
Periventricular leukomalacia in survivors	3 (3.2)	2 (1.5)	3 (8.8)	.11
Necrotizing enterocolitis in survivors	1 (1.0)	6 (4.5)	0 (0)	.20
Respiratory neonatal outcomes				
Respiratory distress syndrome	28 (29.2)	62 (45.9)	21 (61.7)	<.01*
Need for ventilatory support in survivors	61 (63.5)	103 (76.3)	31 (91.2)	<.01 [†]
Duration ventilation in those needing (d)	4	5	6	

Data are n, n (%), or mean±standard deviation unless otherwise specified.

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ISSN: 0029-7844/14



TABLE 1. Fetal Death Rates With Inpatient and Outpatient Care, Studies Published Since 2005

Study	Fetal Deaths, Outpatient (N)	Total Outpatient Fetuses (N)	Fetal Deaths, Inpatient (N)	Total Inpatient Fetuses (N)	Notes
Heyborne ¹⁹	13	88	0	86	Table V
Ezra ³¹	N/A	N/A	0	20	Outpatient cohort excluded (see text)
Defalco ²⁰	3	24	0	22	Table 1 and 2
Murata et al ³³	0	2	1	52	
Van Mieghem ³²	5	106	2	142	Table 1
Quinn et al ⁸	0	0	0	34	
Pasquini ⁹	0	40	N/A	N/A	
Totals	21	260	3	356	
Death rate (%)	8.1		0.8		P < 0.0001

Post. Managing Monoamniotic Twin Pregnancies. CLINICAL OBSTETRICS AND GYNECOLOGY Volume 58, Number 3, 643–653 2015.



Ultrasound Obstet Gynecol 2006; 28: 681–687
Published online in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/uog.3811

High perinatal survival in monoamniotic twins managed by prophylactic sulindac, intensive ultrasound surveillance, and Cesarean delivery at 32 weeks' gestation

L. PASQUINI, R. C. WIMALASUNDERA, A. FICHERA, O. BARIGYE, L. CHAPPELL and N. M. FISK Centre for Fetal Care, Queen Charlotte's and Chelsea Hospital & Institute of Reproductive and Developmental Biology, Imperial College London, Hammersmith Campus, London, UK

- 40 embarazos sobre 20 semanas.
- Sulindac 200 mg para mantener ILA 5-8 cm.
- Us semanal para evaluar ILA, IP AU. Velocidad sistolica de DA c/4 semanas. Interrupción por cesárea a las 32 semanas.
- Reducción 40% ILA, Sobrevida 100%.



- Riesgo de PP vs Óbito fetal.
- La tasa de óbito fetal acumulada es de 0.85% a 1.8% entre las 26 a 34 semanas. Se pueden extrapolar una tasa de óbito semanal no mayor de 2/1000 por semana¹.
- Algunos reportes de 4% mortalidad perinatal sobre las 32 semanas (incluyendo anomalías fetales, STFF y complicaciones por PP).

^{1.} Post. Managing Monoamniotic Twin Pregnancies. CLINICAL OBSTETRICS AND GYNECOLOGY Volume 58, Number 3, 643–653 2015.





The Journal of Maternal-Fetal and Neonatal Medicine 2003;13:414-421

Perinatal outcomes in monoamniotic gestations

H. Roqué¹, J. Gillen-Goldstein², E. Funai³, B. K. Young² and C. J. Lockwood³

133 embarazos

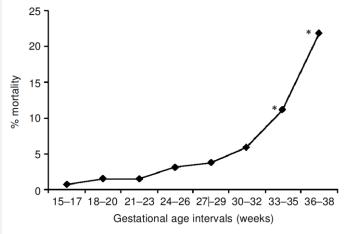


Figure 3 Per cent fetal/neonatal mortality per gestational age interval. *, p < 0.05 compared to gestational age interval 30–32 weeks

Post. Managing Monoamniotic Twin Pregnancies. CLINICAL OBSTETRICS AND GYNECOLOGY Volume 58, Number 3, 643–653 2015.

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Cociliane Database of Systematic Reviews

Planned early delivery versus expectant management for monoamniotic twins (Review)

Shub A, Walker SP

- 2015
- Comparar entre interrupción temprana planeada a las 32-34 semanas vs manejo expectante hasta 36-38 semanas.



Data collection and analysis

No trials were identified by the search strategy.

Main results

No trials were identified by the search strategy.

 Conclusiones: Gemelos monoamnióticos son raros, no hay suficiente evidencia randomizada controlada para determinar el mejor manejo. En ausencia, debemos referirnos a serie de casos y opinión de expertos. El manejo debe tener estas consideraciones. La paciente y su familia debe estar involucrado en esta decisión.



• La escasa evidencia y opinión de expertos sugiere interrupción tipicamente entre 32-34 semanas.

 No existen estudios sobre la vía de parto, se sugiere cesárea por el potencial riesgo de compresión de cordón.

¿Cómo se ha manejado?



			•							
Author	N	Units	Anomalies	тттѕ	Fetal surveillance setting	Delivery timing	Fetal death	NND	Total PMR	PMR nonanomalies
Hack et al. [5]	98	10	7/98 (7%)	6/98 (6%)	Inpatient, 30–32 weeks, in four units	Around 32–34 weeks	22/196 (11%)	12/196 (6%)	34/184 (19%) ≥20 weeks	28/170 (17%) ≥20 weeks
Dias et al. [6]	18	1	2/18 (11%)	0	Unspecified	Around 34 weeks	2/36 (6%)	2/36 (6%)	4/36 (11%) ≥16 weeks	2/34 (6%) ≥16 week
Baxi and Walsh [15]	25	1	9/50 (18%)	1/25 (4%)	Inpatient, ≥26–28 weeks	By 34 weeks	1/50 (2%)	3/50 (6%)	4/50 (8%) ≥20 weeks	$1/41 (2\%) \ge 20$ week
Murata et al. [7]	38	7	Excluded	NA	Inpatient, ≥24–26 weeks	Around 32–34 weeks	17/76 (22%)	0	1/60 (2%) ≥22 weeks	1/60 (2%) ≥22 week
Van Mieghem et al. [8*]	193	8	53/386 (14%)	5/193 (3%)	Inpatient, ≥26-28 weeks, in four units	Around 32–34 weeks	70/386 (18%)	17/386 (4%)	87/386 (23%) ≥11 weeks	60/333 (18%) ≥11 weeks

N, number of cases of monoamniotic twin pregnancies; NND, neonatal death; PMR, perinatal mortality rate at 28 days of age; TTTS, twin-twin transfusion syndrome; units, number of participating centers; weeks, weeks of gestation.

Ishii. Prenatal diagnosis and management of monoamniotic twins. Copyright © 2015 Wolters Kluwer Health.

¿Cómo se ha manejado?

Table 3. Characteristics of the studies included in the meta-analysis. MCMA=monochorionic monoamniotic; IUFD= Intra Uterine Fetal Death; LB=live birth; NNM= neonatal death; CS=caesarean section. *One triplet case excluded.

Author, year	Study period	Study population	Study design	Country	Antenatal management from second trimester	IUFD	LB	NND	Strobe Score
Allen, 2001	1993-2000	25	Multicentric, retrospective	Canada	Unclear	1	41	0	12/18
Sau, 2003	1994-2000	7	Single centre, retrospective	U.K.	US every 2 weks, CS at 32 weeks	1	7	1	12/18
Demaria, 2004	1993-2001	19	Single centre, retrospective	France	US every 2 weeks, CS at 36 weeks	5	25	3	14/18
Heyborne, 2005	1993-2003	96	Multicentric, retrospective	U.S.	Unclear	13	163	2	14/18
Cordero, 2006	1990-2005	36	Single centre, retrospective	U.S.	According to subgroup, CS at 32-34 weeks	0	60	1	14/18
De Falco, 2006	1991-2001	26	Multicentric, retrospective	U.S.	According to subgroup	3	40	0	14/18
Pasquini, 2006	1994-2005	43*	Single centre, retrospective	U.K.	US every 4 weeks, CS at 34 weeks	0	40	0	13/18
Hack, 2009	2000-2007	103	Multicentric, retrospective	Netherlands	Admission at 30-32, CS at 32-34 weeks	5	164	5	13/18
Baxi, 2010	2001-2009	25	Single centre, retrospective	U.S.	Admission at 26-28, CS at 34 weeks	0	41	1	12/18
Dias, 2010	2001-2008	32	Single centre, retrospective	U.K.	US every 4 weeks, CS at 34 weeks	0	32	0	16/18
Morikawa, 2012	2002-2009	101	Multicentric, retrospective	Japan	Unclear	12	163	2	14/18
Murata, 2013	2001-2011	38	Single centre, retrospective	Japan	Admission at 24-26, CS at 34 weeks	1	53	3	14/18
Suzuki, 2013	Unclear	18	Single centre, retrospective	Japan	Unclear	4	23	0	11/18
Van Mieghem, 2014	2003-2012	117	Multicentric, restrospective	Belgium, Canada, Netherlands, Switzerland	According to individual centre's protocol	8	223	8	14/18
Prefumo, 2014	2004-2013	10	Single centre, retrospective	Italy	Admission at 28 weeks, CS at 32 weeks	2	18	0	Not assessed

Prefumo. The natural history of monoamniotic twin pregnancies: a case series and systematic review of the literature. 2015

Conclusiones



- Gemelos monoamnióticos son poco frecuentes, alrededor de 600 casos publicados desde los años 90.
- No hay evidencia randomizada controlada para determinar manejo más adecuado de estas pacientes (500/500).
- En su ausencia, debemos referirnos a serie de casos y opinión de expertos.

Conclusiones



- Por lo que nuestro manejo debe tener estas consideraciones.
- La paciente y su familia tienen que estar involucrados en el proceso y en la decisión de interrupción, explicándoles los riesgos de prematurez vs mortalidad perinatal propia de la patología.





