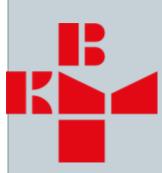
# Prenatal Diagnosis and Counseling on Myelomeningocele in Croatia



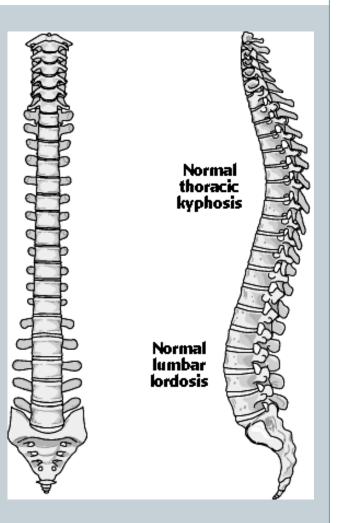
RATKO MATIJEVIC UNIVERSITY OF ZAGREB, SCHOOL OF MEDICINE MERKUR HOSPITAL ZAGREB, CROATIA



# Normal spine

# Visualize (and document):US and MRI

- Cervical, thoracic, abdominal, lumbar and sacral part
- Transverse and longitudinal view
- Triangular shape of osification centres (closed triangle)



# MRI

 increasingly used in clinical practice, partly because of the increasing interest in fetal surgery and fetal medicine

#### Ultra-fast MRI

Glenn OA and Coakley FV. MRI of the fetal central nervous system and body. Clin Perinatl 2009;36:273-300



# Ultrasound examination (level 1) 018-22 weeks • 12-14 weeks

# Longitudinal









# Spina bifida – type of myelomeningocele

#### Neural tube defect

- o Forms 5 6 weeks
- o Lumbar and sacrolumbar the most frequent
- Incidence 0.5-2:1000 (Croatia 0.5/1000 livebirth)
- o Related to chromosomal anomalies

#### Prognosis dependant on extent of the lesion

Possible total paralysis below the lesion

#### Related to:

- o Hidrocephalus
- o Congenitalnital defecs
- Meckel's syndrom (polidactily, multicystic kidneys autosomal recessive)
- o Amnotic band sindrome (not genetic)

#### Classification

#### Closed

Covered by skin Normal AFP





#### Open

Not covered by skin, drainage of the cerebrospinal liquor
 Reject AER

- o Raised AFP
- Rachishisis: complete open spine with herniation and/or destruction of neural spine





#### Cystic

• Meningocoele or meningomyelocoele (myelomeningocoele)



#### Ocult – not type of myelomeningocoele

Part of the vertebrae is not closed but without open defect
No clinical significance and without raised AFP



# Dijagnosis

#### • US

- Transversal plane the best for diagnosis but sagital better to assess the extent of the lesion
- Protrusion of the sac (meninges) with spinal cord (or not)
- Indirect signs (banana and/or lemon) – obliterated cisterna magna



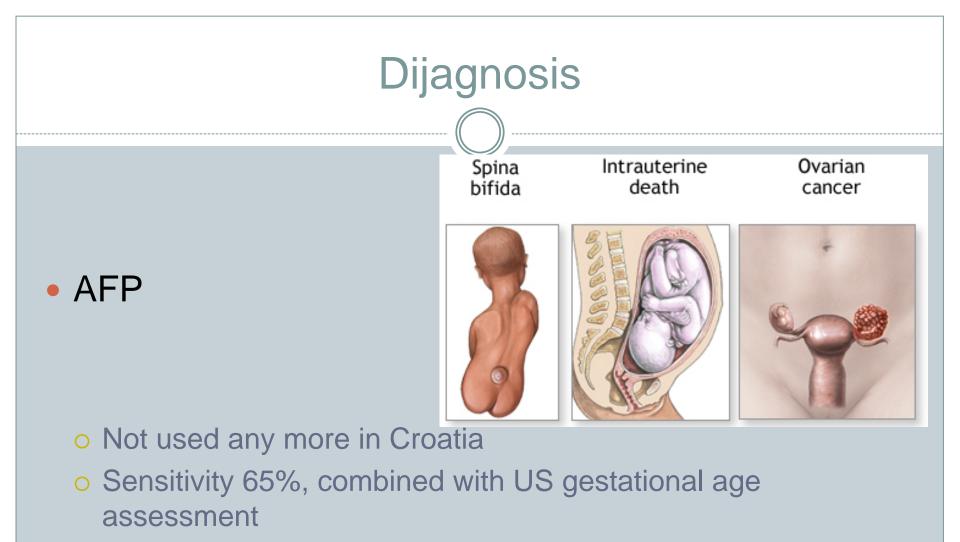
# Diagnosis

#### • MRI

o Since 2000

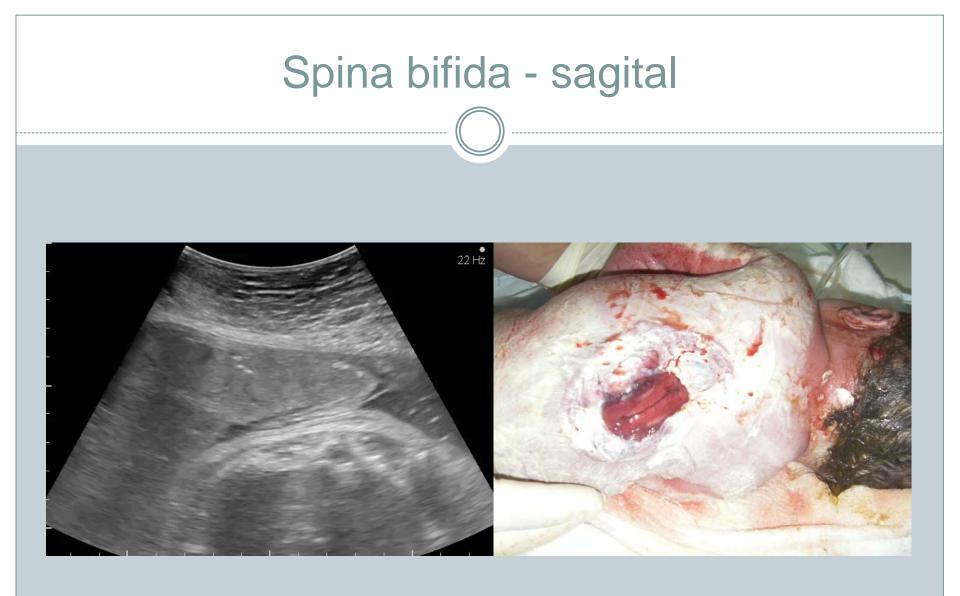
- Rarely used in Croatia
- Helpful..., problems in interperetation of immages
  Compared to US





- US sensitivity up to 100% (P< .001 compared with screening) but later in pregancy
  - × Dashe J i sur. AJOG 2006;195:1623





#### Associated anomalies

- Hidrocephalus, encephalocolela-e, Arnold Chiari
- Meningomielocoele (more frequent that meningocoele)
- Any level on fetal spine
- Abnormal osification of posterior centres

### Ultrasound diagnosis - Croatia

- Incidence 0.5/1000 (livebirth) lower rate
- Diagnostic accuracy 50-100% (depending on centre)
- TOP if before 22/40
- Folic acid uptake different

#### Referal centre

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61         VSD         7         1         14           62         VSD         7         1         14           63         Ventriculomegalia         8         8         100           64         Omphalocele         8         8         100           65         Cystic hygroma         10         8         80           66         Ovarian cyst         11         11         100           67         Hydrocephalia         11         11         100           68         Gastroshisis         11         11         100           69         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multiple malformations         13         11         85           70         Polydactily unilateral         24         1         4           71         Hydronephrosis unilateral         32         2         6           74         Clubfoot Talipes equinovarus unilateral         32         2         6	59		6	0	0	
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63         Ventriculomegalia         8         8         100           64         Omphalocele         8         8         100           65         Cystic hygroma         10         8         80           65         Ovarian cyst         11         11         100           67         Hydrocephalia         11         11         100           67         Hydrocephalia         11         11         100           69         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multiple malformations         13         11         85           72         Polydactily unilateral         24         1         4           74         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76         76         76         76	61					
64         Omphalocele         8         8         100           65         Cystic hygroma         10         8         80           65         Cystic hygroma         11         11         100           66         Ovarian cyst         11         11         100           67         Hydrocephalia         11         11         100           68         Gastroshisis         11         11         100           69         Multicystic dysplatic kidney unilateral         12         11         92           70         Cleft lip and palate         13         11         85           71         Multiple malformations         13         11         85           72         Polydactily unilateral         24         1         4           74         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76           6	62					
65         Cystic hygroma         10         8         80           66         Ovarian cyst         11         11         100           7         Hydrocephalia         11         11         100           68         Gastroshisis         11         11         100           69         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multicystic dysplastic kidney unilateral         24         1         4           72         Polydactily unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76           4	63					
66         Ovarian cyst         11         11         10           67         Hydrocephalia         11         10         91           68         Gastroshisis         11         10         91           68         Gastroshisis         11         11         100           9         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multicyle malformations         13         11         85           72         Polydactily unilateral         24         1         4           74         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76         76         76         76	64		_			
67         Hydrocephalia         11         10         91           68         Gastroshisis         11         11         100           69         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multiple malformations         13         11         85           72         Polydactily unilateral         24         1         4           73         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           76	65					
68         Gastroshisis         11         11         100           69         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multicystic dysplastic kidney unilateral         13         11         85           72         Polydactily unilateral         24         1         4           73         Hydronephrosis unilateral         28         6         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76           6						
69         Multicystic dysplastic kidney unilateral         12         11         92           70         Cleft lip and palate         13         3         23           71         Multiple malformations         13         11         85           72         Polydactily unilateral         24         1         4           73         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76           4						
70         Cleft lip and palate         13         3         23           71         Multiple malformations         13         11         85           72         Polydactily unilateral         24         1         4           73         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76         76         76         77						
71         Multiple malformations         13         11         85           72         Polydactily unilateral         24         1         4           73         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76						
72         Polydactily unilateral         24         1         4           73         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76         76         75         76						
73         Hydronephrosis unilateral         28         26         93           74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76         76         76         76						
74         Clubfoot Talipes equinovarus unilateral         32         2         6           75         76	73			-		
75 76	74					
	75			-	-	
	76					

#### • Folic?

o Vit B

Reduces the risk up to 70% - BEFORE PREGNANCY

#### Natural sources

- Vegetables, fruit, juices
- o Insufficient

#### • How much?

- o 400 mcg (0.4 mg)
- High risk (history) 4000 mcg (4.0 mg) three months

# **Ireland example**

- 225 998 births, 236 NTD, 1.04/1 000
- 0.92/1 000 in 2009 to 1.17/1 000 in 2011
  - o 45% (n = 106) had an encephaly
  - o 49% (n = 115) had spina bifida
  - o 6% (n = 15) had an encephalocoele
- Peri-conceptional folic acid supplement intake was 13.7% among the 52.5% (n = 124) of cases whose folic acid supplement intake was known.
- ?mandatory folic acid food fortification

McDonnel R et al. Neural tube dfects in the Republic of Ireland 2009-2011.J Public Health 2014

# Something new?

european surveillance of congenital anomalies

 Neural tube defects (NTDs) have not significantly decreased in prevalence over the last 10 years, showing that efforts to prevent NTD with periconceptional folic acid supplementation have been largely unsuccessful.

Newsletter, 22nd december 2014

# Antiepileptics

- o Valporate, filate use, increasing the dose
- Jentink et al. Does folic acid use decrease the risk for spina bifida after in utero exposure to valproic acid? Pharmacoepidemiolog Drug Saf 2010;19:803
  - o OR with folic acid
    - not exponated0.5 [95%CI: 0.3-0.7]eksponated1.0 [95%CI: 0.1-7.6].
- Conclusion not possible
- Folic acid may not reduce the risk of spina bifida realted to valoprate use, but reduces the risk in general

#### Inconclusive regarding the dose

Croatia: "Why not?" principle, 10 times the normal dose



www.nih.gov 2014 Adzik NS. Sem Pediat Sur, 2013



#### LABOR AND DELIVERY

CAESAREAN SECTION IS THE COMMONEST WAY, mandatory if breech, possible if there is no makrocephaly
Wilson RD et al J Obstett Gynaecol Can 2014;36:927
Is Cesarean helpful regarding neonatal outcome – remains open question
Hill AE and Beattie F. Eur J Paediatr Surg 1994;4:32
No benefit of Cesarean in there is no other indication
McLone DG and Bowman RM. Up-to-Date 2014

Croatia: Common sense... (menaing Caesarean)



#### Post natal manegement Croatia

- Postdelivery care O & G
- Postnatal surgery 5 centres
- On average 20 year (+/-3) HALFED IN RECENT YEARS
- Postnatal transfer
  - Graz, Austria

(rare)



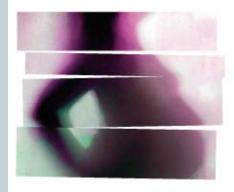
#### Colaboration







# Thank you



6<sup>th</sup> Congress of the South-East European Society of Perinatal Medicine



Zagreb / CROATIA 4-6. 12. 2015.

The South East European Society of Perinatal Medicine