Diagnostic workup, patient selection and preoperative management – The Zurich approach

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4 ethical principles

Autonomy

(voluntas aegroti suprema lex)

Beneficence

(salus aegroti suprema lex)

Non-maleficence

(primum nil nocere)

Justice

Related to spina bifida repair

Autonomy

- Non-directive counselling

Beneficence

- Reversal of hindbrain herniation preventing shunts
- Improvement of peripheral neural function

Non-maleficence

- Prevention of preterm birth
- Preservation of maternal reproductive health

Justice

- Accepting of all patients
- Treatment in experienced centers with long term follow-up

Patient selection

Inclusion Criteria

- Singleton pregnancy
- Isolated spina bifida T1-S1
- Evidence of hind brain herniation
- Gestational age 19 0/7-25 6/7 weeks
- Normal karyotype
- Maternal age ≥18 years

Exclusion Criteria

- Fetal anomaly unrelated to spina bifida
- Severe kyphosis
- Preterm birth risk (short cervix, history)
- Placental abruption
- BMI >35
- Contraindication to surgery including previous hysterotomy in the active uterine segment

Preoperative counselling

- Multiple counselling sessions with obstetrician and pediatric surgeon
- Teaching about neural tube defects and their long term consequences
- Strength, Weakness, Opportunities and Treats of fetal surgery and alternatives (SWOT)
- Hooking patients with parents of affected children (both with fetal and neonatal repair)

Diagnostic workup

Diagnostic workup - mother

- Obstetric and general history
- Cervical length (scan)
- Fibronectin
- Screening for vaginal and urinary bacterial infection
- Serological testing for HIV, HBV, HCV
- Blood group, irregular antibodies
- ECG

Diagnostic workup - Fetus

- Detailed ultrasound scan including
  - Ventricles
  - Hindbrain
  - Shape of the spine
  - Dimension of the spina bifida lesion
  - Position of feet
  - Leg, foot and toe movements
  - Position of placenta and cord insertion
- Fetal MRI
- Fetal karyotype and high resolution array

Associated syndromes with spina bifida

Adams-Oliver Syndrome (Romani et al,1998)
Antley-Bixler Syndrome (Chun et al,1998)
Asplenia Syndrome (Van Went et al,1977)
Becker Naevus Syndrome (Happle and Koopman,1997)
Down's Syndrome (Gal,1971; Szabo et al,1986) ←
Dubowitz Syndrome (Hansen et al,1995)
Edward's Syndrome (Trisomy 18) ←
Gorlin Syndrome (Ratcliffe et al,1995)
Jarcho-Levin Syndrome (Giacoia and Say,1991)
Klippel-Feil Syndrome (Jablonski,1969)
Larsen Syndrome (Anderson,1997)
Marfan's Syndrome (Ortino et al,1988)
Mayer-Rokitansky-Kuster-Hauser Syndrome (Strubbe et al,1992)
Neu-Laxova Syndrome (Naveed et al,1990)
Patau Syndrome (Rodriguez et al,1990) ←
13q- Syndrome (Chemke et al,1978)
VATER Syndrome (Quan and Smith,1973)
Velo-cardio-facial Syndrome (Nickel et al, 1996)
Waardenburg Syndrome (De Saxe et al,1984)
XX-Agonadism Syndrome (Kennerkrecht et al,1997)

Reason for rejection	n
Twin pregnancy	1
Level S2	1
Gestational age > 25 6/7	4
Abnomal karyotype, anomalies	4
BMI >35	1
Maternal non-compliance	1
MMC repair declined by patients	12

Preop management

OP – 2d

- Written informed consent
- Steroids for lung maturation if >24 weeks

OP – 1d

- Tested packed red cells (0 neg) for fetal transfusion if needed
- Omeprazol 40mg p.o.

OP – 1h

- Indomethacin 50mg rectal
- Cervical length measurement
- Cephazolin 1g i.v.