Neonatale aanpak van het kind met een anorectale malformatie

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Anorectal malformations

- **Incidence:** 1/4000-5000 live births
- **Occuring ≤ 6th-7th week of gestation**
- **Very heterogenous group of severity**
  - Anatomy of the malformation
    - high vs low: supralevator vs infralevator
    - +/- fistula = ectopic anal canal (IAS?)
  - Associated anomalies (2/3)
    - chromosomal defects: 8%
- **Pelvic floor function**
  - anatomy
  - innervation
Normal anatomy

Stec, Semin Pediatr Surg 2011
Normal anatomy

PC line and I point = levator complex
Determinants of fecal continence in ARM

- Type of malformation
- Associated anomalies
- Surgical aspects
- Postoperative care
Determinants of fecal continence in ARM

- Anatomical sphincter defects
- Decreased/absent anorectal sensitivity
- Motility disorders
- Chronic rectal distention
- Lumbosacral nerve abnormalities
Which classification for ARM

• **Many classifications – difficult comparison**
  
  • Ladd and Gross 1934
  • Stephens and Smith 1963 (high vs. low)
  • Santulli 1964
  • “International” Melbourne Classification 1970 (nearly 40 subtypes)
  • Wingspread Classification 1984
  • Peña 1995
Wingspread Classification

- Fistula vs. no fistula
- Low vs high vs intermediate
To develop a system for comparable follow-up studies
International classification for anorectal malformations (Krickenbeck)
Holschneider et al, J Pediatr Surg 2005

Major clinical groups

- All:
  - No fistula
  - Anal stenosis
  - Perineal (cutaneous) fistula
  - Rectovesical fistula

- Female:
  - Vestibular fistula
  - Cloaca

- Male: Rectourethral fistula
  - Bulbar fistula
  - Prostatic fistula

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- Rare/regional variants
  - Pouch colon
  - Rectal atresia/stenosis
  - Rectovaginal fistula
  - H type fistula
  - Others
International classification for anorectal malformations (Krickenbeck)

Holschneider et al, J Pediatr Surg 2005

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> 50%
Perineal cutaneous fistula

• = covered anus, incomplete (Int. Melbourne Classification)

• Commonest male anomaly
Perineal cutaneous fistula

“White epithelial pearl”

Black ribbon
(Anterior) ectopic anus

- Abnormally located anus, mainly in girls
  - Girls: anal index < 0.34
  - Boys: anal index < 0.46

Reisner et al, Pediatrics 1984

Surrounded by internal and external sphincter

Problem = Constipation

In most cases no surgery needed


Anal index = \[
\frac{\text{scrotal or fourchette/mid anal distance}}{\text{Mid anal distance /coccygeal distance}}
\]
• Common wall between rectum and vagina
Rectourethral fistula

- Bulbar fistula more common than prostatic
- Common wall between fistula and urethra
International classification for anorectal malformations (Krickenbeck)

Holschneider et al, J Pediatr Surg 2005

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The other major clinical groups
Anal stenosis

- **Covered anus:**
  - (Partial) covering by genital folds
    (hypertrophic raphe, bucket handle)

- **Anal membrane**
Rectovesical fistula

- rectobladder neck fistula
- Flat perineum
- Sacrum and pelvis can appear dysmorphic
Associated anomalies (1)

- More associated anomalies in high lesions
- Serious, potentially lethal defects
- Vertebral - spinal deformities
  - hemivertebrae
  - sacral deformities
  - caudal regression syndrome
  - tethered cord?
    - skin lesion: lump, vascular nevus, sinus, angioma on the midline of the back, hypertrichosis, skin dimple, sacral lipoma
    - neurological or neuro-orthopedic abnormalities of lower extremities, scoliosis
    - bladder and bowel dysfunction

Neural tube defect of terminal spinal cord:
- Vertebral abnormalities
- Flat buttocks
- Lower limb neurological deficit
- Neurogenic bladder
- ARM

permanent colostomy
Associated anomalies (2)

- **Genitourinary**
  - in 50% of cases
  - genital, urethra, bladder, ureter, kidney

- **VACTERL association**
  - esophageal atresia
to be ruled out

- **Other intestinal malformations**
  - Hirschsprung’s disease = rare

- **Trisomy 21**: mostly no fistula – good prognosis

Initial assessment of the newborn

• Clinical assessment
  – Perineal inspection
  – Associated anomalies

• Imaging
  – Cross-table lateral film
  – Ultrasound, cystogram, spinal examination
Perineal inspection

- Anal dimple = EAS (cutaneoanal reflex, EMG)
- Fistula visible?
- Midline groove between buttocks and anal dimple ("flat buttocks")
  decreased prominence ~ height of the fistula
- Boys:
  - meconium or squamous epithelium in urine = fistula
  - white epithelial pearls = fistula
- Girls: number of orifices + probing
Prone cross-table lateral X-ray

- After 24 hours!
- Wait 3 minutes in this position
- Accurate centering on greater trochanter
- Cave meconium plugs, crying, levator muscle contraction

Narasimharao et al, AJR 1983
Voiding cystourethrography

- Antegrade urethrogram + retrograde whilst removing the catheter

- In all patients
  - Renal tract anomalies?
  - Fistula? (but less reliable than colostogram)
Imaging

- Prone cross-table lateral film
- Cystourethrography
- Ultrasound abdomen
  - Kidney, pelvis (genitourinary)
- Transperineal ultrasonography for location of distal rectal pouch (> 15 vs. < 15mm) and fistula Kim et al, 2000; Haber et al, 2007
- Echocardiography
- Spine
  - X-ray spine
  - X-ray pelvis-sacrum
  - US spine for detection of spinal dysraphism
  - MR spine
Sacral ratio

Normal sacral ratio: $a/b \geq 0.74$ (anteroposterior)

If sacral ratio < 0.5: bad prognostic sign for ultimate continence
Routine spinal MR imaging in all patients?
Alamo et al, Radiographics 2013
Management of the newborn - male

Perineal inspection

- Spine
- Kidney U/S
- Urinalysis
- R/O esophageal atresia
- Sacrum
- Spinal U/S
- Cardiac echo

Re-evaluation and cross-table lateral film

- Perineal fistula
  - Anoplasty
- Rectal gas below coccyx
  - No associated defects
    - Consider with or without colostomy
- Rectal gas above coccyx
  - Associated defects
    - Abnormal sacrum
      - Flat bottom
      - Colostomy

20 - 24 hrs

micturating cystourethrogram

final treatment

Levitt and Peña, Curr Opin Pediatr 2005
Perineal repair - male

Cutback anoplasty
Management of the newborn - female

Levitt and Peña, Curr Opin Pediatr 2005
Colostomy formation

Later high-pressure colostogram
International grouping of surgical procedures (Krickenbeck)

- **Pull-through abdominoperineal** (Rhoads, 1948)
- **Sacroperineal approach** (Stephens 1953)
- **Perineal operation** (Browne, Potts 1954)
  - Cutback anoplasty (Browne)
  - “Potts transfer anoplasty”
- **Pull-through abdominosacroperineal** (Kiesewetter 1966, Rehbein 1967)
- **PSARP** (Peña-devries, 1982)
- **Anterior sagittal approach** (Mollard, 1989, Okada, 1992)
- **Pull-through laparoscopic-assisted** (Georgeson, 2000)

Holschneider et al, J Pediatr Surg 2005
PSARP
Low ARM: histopathology of distal 3cm
Lombardi et al, J Pediatr Surg 2013

95% of good functional results without soiling or constipation (FU 3-8 years)

No IAS structure recognised

avoid minimal dissection and minimal resection of fistula?
### Indicators of prognosis for bowel control in ARM patients

<table>
<thead>
<tr>
<th>Good prognostic features</th>
<th>Poor prognostic features</th>
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<tbody>
<tr>
<td>• Normal sacrum / spine</td>
<td>• Abnormal sacrum</td>
</tr>
<tr>
<td>• No presacral mass</td>
<td>• Myelomeningocele</td>
</tr>
<tr>
<td>• Good buttock crease</td>
<td>• Some types of ARM</td>
</tr>
<tr>
<td>• Good anal dimple</td>
<td>• Rectoprostatic urethral fistula</td>
</tr>
<tr>
<td>• Some types of ARM</td>
<td>• Rectobladder neck fistula</td>
</tr>
<tr>
<td>◦ Rectoperineal fistula</td>
<td>• Cloacal extrophy cloaca</td>
</tr>
<tr>
<td>◦ Rectobulbar urethral fistula</td>
<td>• &gt;3 cm common channel</td>
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<tr>
<td>◦ Rectovestibular fistula</td>
<td>◦ Complex defects</td>
</tr>
<tr>
<td>◦ Cloaca &lt; 3 cm common channel</td>
<td></td>
</tr>
<tr>
<td>◦ Rectal atresia or stenosis</td>
<td></td>
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<tr>
<td>◦ Imperforate anus without fistula</td>
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Levitt et al, J Pediatr Surg 2010
Conclusions

• A good, simple classification is crucial for adequate comparison of data
  • Surgical and prognostic relevance
  • “high and low” is too general
  • Ectopic anus is not the same as ectopic anal canal

• Multidisciplinary collaboration
  • Pediatrician, pediatric surgeon, radiologist, nursing team, psychologist, physiotherapy, stoma nurse…
    • Preoperative work-up
    • Meticulous surgical technique
    • Postoperative follow-up into adulthood
      • GI, urinary, sexual, psychosocial
THANKS!