Quality of Life of Patients with an Anorectal Malformation or Hirschsprung Disease and their Parents

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Curriculum Vitae

- Born 2nd October 1980, Hoorn, The Netherlands

- ’99 - ’06 Medical School, University of Amsterdam,
- ’06 - ’08 Medical doctor (The Hague, Zaandam)
- ‘08 – ’14 Surgical resident, VUMC, Amsterdam
- ‘14 – ’16 Fellow Pediatric Surgery, Amsterdam and Groningen
- ‘16 - …. Pediatric Surgeon

- ‘12 - ….. PhD project: KLANKbord-study




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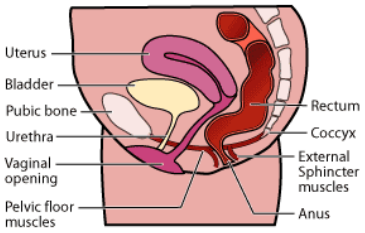
Introduction

- Incidence ARM worldwide 1 in 1000-5000 living births
- Approximately 40 new cases a year in the Netherlands

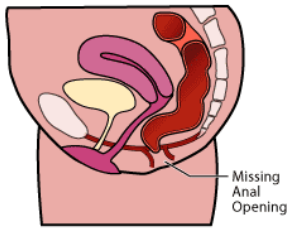
- Fecal incontinence, constipation and soiling are some of the problems that can be encountered during life.
- Do these problems influence the QoL of your patient with ARM?

Anatomy

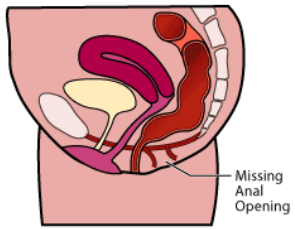
Normal Anatomy Female



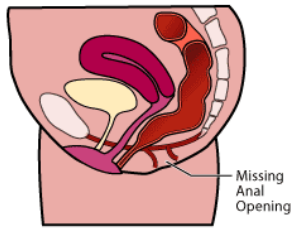
Anal opening missing or closed off



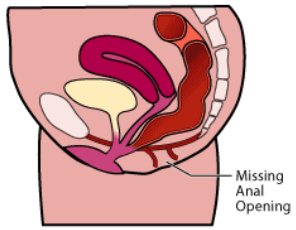
Opening in wrong place and too small



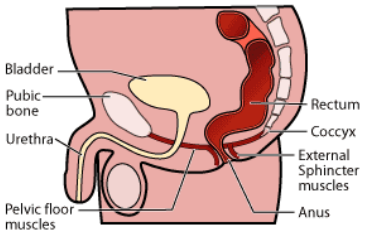
Rectum connects to vagina



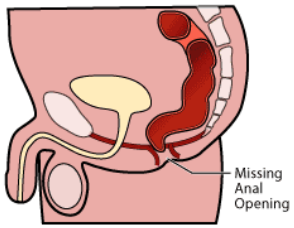
Cloaca: all three form a single opening



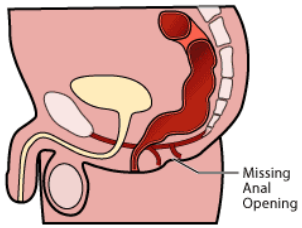
Normal Anatomy Male



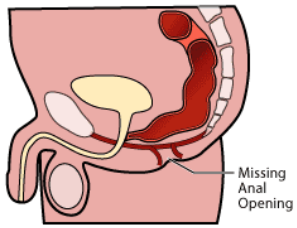
Anal opening missing or closed off



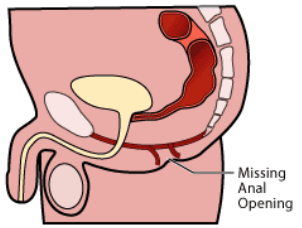
Opening in wrong place and too small



Rectum connects to urethra or bladder



High rectum connects into bladder



What is QoL?

Quality of Life:

"Individual's perception of his/her position in life in the context of the culture and value system in which he/she lives and in relation to his/her goals, expectations, standards and concerns (Physical, psychological, social, environment, level of independence and spirituality)"

→ Subjective and refers to satisfaction

Health Related Quality of Life:

Those aspects of life that are directly influenced by one's health

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BAPS Paper

Qualitative analysis of studies concerning quality of life in children and adults with anorectal malformations

Marieke J. Witvliet[1], Annelie Slaar[2], Hugo A. Heij, Alida F.W. van der Steeg[3]

Material and Methods - 1

- Literature search
  - Pubmed (118)
  - Psychinfo (22)
  - Cochrane Library (0)
  - Web of Science (100)

- “Quality of Life” ; “Anal Atresia” ; “Anorectal Malformation” ; “Imperforate anus”

Material and Methods - 2

- Selection criteria:
  - Studies concerning patients with ARM
  - QoL is measured with questionnaires
  - Article published in peer-reviewed journals
  - Published in English or Dutch

→ 30 articles met all criteria and were included

Material and Methods - 3

- Validated QoL questionnaires:
  - TACQoL
  - PedQl
  - Kidscreen
  - CHQ/ITQoL
  - DUX
  - SF-36
  - WHOQOL(-Bref)
  - Questionnaire validated in a validation article

QoL assessment:
1. Definition of QoL is provided
2. QoL is assessed with a validated and reliable questionnaire
Study population:
3. A description is given of the initial anomaly
4. A description is given of the surgical treatment
5. A description is given of associated anomalies (VACTERL-association)
6. A description is given of current treatment (bowel-management, medication)
7. Inclusion and/or exclusion criteria are provided
8. Participation rate is > 75%
9. MEC approval and/or informed consent was given
Study design:
10. A power analysis is provided to account for the number needed to include
11. The collection of data is prospective
12. The design is longitudinal and includes more than one measurement moment
13. Multivariate analysis is done
14. The follow-up period is at least 2 years
15. The loss to follow up is < 20%
16. A comparison of participants and non-participants is done
17. A comparison of dropouts and non-dropouts is done
18. In case of a subgroup, the characteristics of the subgroup are clearly described
Results:
19. The results are compared between 2 groups or more (e.g. healthy population, groups with different intervention therapy, groups with different type of anorectal malformation) and/or results are compared with at least two time points (e.g. longitudinal)
20. Psychometrically sound questionnaires are used to measure QoL and/or functional outcomes

Maximum = 20 points



Material and Methods - 5

- Scoring of the questionnaires:
  - ≥ 14 “High” quality study
  - 10-13 points “Moderate” quality study
  - ≤ 9 “Low” quality study
- Disagreements solved through discussion in consensus meeting

Results - 1

- Quality of the studies:
  - Score range 2 – 13
  - Mean score 6.8 points
  - “Low” quality 26 studies
  - “Moderate” quality 4 studies
  - “High” quality 0 studies

Results - 2

- Study characteristics:
  - All cohort studies
  - Study size:
    - <50 pts 12 studies
    - 50-100 pts 8 studies
    - 100-200 pts 3 studies
    - >200 pts 7 studies
  - 4 studies longitudinal and >1 measurement
  - 28 studies described the original anomaly
  - 13 studies described the surgical treatment
  - 6 authors used validated QoL questionnaires

Results - 3

- Studies considering adults (n=9)
  - QoL was affected by fecal incontinence in 3 studies (no validated questionnaires)
  - 2 studies did not establish a correlation between QoL and fecal incontinence (1 used validated questionnaires)
  - Lower QoL in female patients, patients with other congenital anomalies or with a stoma
  - Self-efficiacy is correlated to body image and sexual functioning

Results - 4

- Studies considering children and/or adolescents (n=21)
  - 9 studies found QoL, psychosocal functioning and depression influenced by fecal incontinence (no validated QoL questionnaires
  - 5 studies stated that their was no correlation (2 used validated questionnaires)

Results - 5

- Fecal continence correlated to QoL:
  - 19 studies
    - 12 studies found a correlation
      - 0 studies used validated questionnaires
    - 7 studies found no correlation
      - 3 studies used validated questionnaires
- 5 studies gave a definition of QoL

Discussion

- A definition of QoL is essential
- Conclusions were drawn without validated QoL questionnaires. Often this was at best health-related QoL
- Operation techniques have changed, but were not always mentioned
- QoL can be influenced by many factors and life events
- Therefore it is a longitudinal changing concept

KLANKbord-study

- Quality of Life (QoL) Analysis in Children and their Parents with an Anorectal Malformation (ARM) or Hirschsprung Disease (HD).
- Started in January 2012
- 3 Pediatric Surgical Centers in the Netherlands, and 2 more starting this year (2016)

Hypothesis

- QoL of children with ARM or HD is dependent of functional problems and is lower than their healthy peers.
- Fear and neuroticism of parents after the diagnosis ARM or HD was confirmed can lead to more parental stress
- High scores on parental stress and/or fear and neuroticism can lead to more functional problems in the child
- More functional problems of the child can lead to a lower QoL

Published articles

Anxiety and Quality of Life of Parents with Children Diagnosed with an Anorectal Malformation or Hirschsprung Disease

Marieke Witvliet[1] Christine Sleeboom[1] Justin de Jong[1] Anton van Dijk[2] Sander Zwaveling[3] Alida van der Steeg[1,4]

- Compared parents of newborns to parents of older children
- Overall (entire group) mothers scored worse than fathers on STAIT anxiety and the psychological domain of the WHOQOL-Bref
- Parents of newborns: STATE anxiety was significantly worse in mothers compared to fathers. TRAIT anxiety and psychological domain showed a trend that mothers scored worse
- Parents of older children: no differences were encountered

Quality of Life and Anxiety in Parents of Children with an Anorectal Malformation or Hirschsprung Disease: The First Year after Diagnosis

Marieke J. Witvliet[1,2] Roel Bakx[1] Sander Zwaveling[3] Tonnis H. van Dijk[2] Alida F. W. van der Steeg[1,4]

- No significant difference in QoL between fathers and mothers and between measurements
- Anxiety levels are significantly higher in mothers than in fathers soon after the diagnosis was made.
- This difference in anxiety levels can not be measured anymore after one year

→ Coping strategies are very important

Projects in progress or submitted

- The transitional outpatient clinic for patients with ARM and HD: What is the effect?
- Do patients with ARM or HD have more sexual problems in later life? And can we predict these problems by patient characteristics in their childhood?
- What is the effect of all the hospital visits on parents of children with ARM or HD using the Pediatric inventory for parents.
- QoL and functional outcomes of the different operations used for long segment HD: Multicenter study in the Netherlands.
- QoL and functional outcomes of the different operations used for short segment HD: Single-center study.

Thank you for your attention