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Long-term follow-up of functional outcome in patients with a cloacal malformation: A systematic review

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Abstract

Background: Reconstructive surgery is performed in patients with cloacal malformations to achieve anorectal, urological, and gynecological function. The aim of this study was to evaluate the functional outcome of cloacal malformation repair as reported in literature.

Methods: A systematic literature search was conducted according to PRISMA guidelines using PubMed, EMbase, and Web-of-Science. Records were assessed for the reporting of functional outcomes, which was divided into anorectal, urological, or gynecological function. Studies were used in qualitative (Rangel score) and quantitative syntheses.

Results: Twelve publications were eligible for inclusion. Voluntary bowel movements were reported in 108 of 188 (57%), soiling in 146 of 205 (71%), and constipation in 31 of 61 patients (51%). Spontaneous voiding was reported for 138 of 299 patients (46%). 141 of 332 patients (42%) used intermittent catheterization, and 53 of 237 patients (22%) had a urinary diversion. Normal menstruations were reported for 25 of 71 patients (35%). Centers with limited experience reported similar outcome compared to centers with more experience (≥ 1 patients/year).

Conclusion: In this review we present functional outcome of the largest pooled cohort of patients with cloacal malformations as reported from 1993 to 2012. Functional disturbances are frequently encountered in anorectal, urological, as well as gynecological systems. Reporting of functional outcome in these patients should improve to increase knowledge about long-term results in patients with this rare malformation and to reach higher study quality. Especially, sacral and spinal anomalies should always be reported given their impact on functional outcome. Specialized care centers may be of great importance for patients with rare and complex conditions.

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A cloacal malformation is one of the most complex congenital anorectal abnormalities and still is one of the most challenging procedures in pediatric surgery. A cloacal malformation is defined as a urethra, vagina, and rectum that remain fused during the early stages of embryological development [1]. Occurring once in every 20,000–25,000 newborns, it accounts for approximately 10% of all anorectal malformations [2,3].

Surgical therapy for cloacas changed considerably with the application of the posterior sagittal approach in 1982 [4,5], and again with the introduction of total urogenital mobilization in 1997 [6]. The reconstruction of cloacal malformations became more anatomically precise, operation time was reduced, and there were fewer postoperative complications [6,7].

In the early years of successful surgical management of cloacal malformations studies focused on short-term results of fecal and urinary continence of the different surgical approaches. In more recent years longer-term fecal and urinary functional outcome of patients with cloacal malformations was reported, as well as reproductive outcomes such as having a normal sexual and reproductive life. However, most studies consisted of small numbers of patients due to the malformation's rarity. Furthermore, many studies include their patients with cloacal malformations as a part of the group of patients with anorectal malformations. Finally, many different scoring systems have been used making comparison of outcomes difficult and associated sacral and spinal anomalies are rarely mentioned. A good overview of the final outcome of all three systems (anorectal, urological, and gynecological) is still lacking. Therefore, a systematic review was performed to analyze all current literature on long-term functional outcome in the three areas that are congenitally malformed by the cloacal malformation.

1. Methods

1.1. Guideline

The PRISMA statement, checklist and flow-chart were used in order to achieve the highest standard in reporting items for a systematic review and meta-analysis [8,9].

1.2. Search strategy

A systematic literature search was conducted on November 11th, 2011 using the PubMed, EMbase, and Web-of-Science databases. Studies were searched in PubMed using the following search terms: (cloaca[mesh] NOT enterobacter [mesh]) OR anus, imperforate[mesh]) AND (surger*[tw] OR surgic*[tw] OR operat*[tw] OR reconstruct*[tw]) AND (outcom*[tw] OR effic*[tw] OR continen*[tw] OR incontinen*[tw] OR soil*[tw] OR catheter*[tw] OR constip*[tw] OR obstipat*[tw] OR menstruat*[tw] OR void*[tw]) NOT (animals[mesh] NOT humans[mesh])) OR ((cloaca*[tw] OR anus*[tw] OR anal[tw] OR anorect*[tw] OR urorectal*[tw] OR rectal*[tw] OR rectum*[tw] OR urogenit*[tw] OR urologic*[tw] OR vagina*[tw]) AND (malform*[tw] OR anomal*[tw] OR imperforat*[tw]) AND (surger*[tw] OR surgic*[tw] OR operat*[tw] OR reconstruct*[tw]) AND (outcom*[tw] OR effic*[tw] OR continen*[tw] OR incontinen*[tw] OR soil*[tw] OR catheter*[tw] OR constip*[tw] OR obstipat*[tw] OR menstruat*[tw] OR void*[tw]) NOT (animals[mesh] NOT humans[mesh]) NOT medline[sb]).

For the other databases similar search terms were applied which concerned the functional outcome of patients operated on for cloacal malformations.

1.3. Eligibility criteria

All written studies in English that reported postoperative functional outcome of patients with a cloacal malformation were included. Functional outcome was defined as anorectal, urological, or gynecological. No limits were set with regard to date of publication. Studies on cloacal exstrophy and the cloacal dysgenesis sequence were excluded, as well as all case-studies or studies presenting less than 5 patients. Studies concerning anorectal malformations in general were only included when presenting at least 5 patients with a cloacal malformation and when results of these patients were reported separately from the results of the patients with other anorectal malformations. The references of each of the articles we found were also reviewed to include useful studies that might have been missed with the initial literature review. Different articles that presented identical variables of the same study population were excluded, and the most recent publication, the publication presenting the largest sample or the most outcome variables was chosen.

1.4. Study selection

The study selection consisted of four separate processes; 1. Study identification, 2. Study screening, 3. Study eligibility, 4. Study inclusion. All processes were conducted by two separate reviewers (HV, IdB).

1.5. Quality assessment

Quality of the articles was scored using the checklist as proposed by Rangel et al. [10]. The checklist consisted of 3 subscales containing 30 items in total. The 3 subscales were: 1. Potential Clinical Relevance, 2. Quality of Study Methodology, and 3. Quality of Discussion and Stated Conclusions. The maximum total score was 45 points. Scores ranging from 0 to 15 indicated a study of poor quality, studies scoring from 16 to 30 points were considered to be fair and scores of 31 points or higher indicated a qualitatively good study. All studies of poor quality (scoring less than 16 points) were excluded.

1.6. Data extraction

Two reviewers (HV, IdB) used predefined criteria to extract the data from included publications. The predefined