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Topic: Females with anorectal malformation (ARM): when is the best time to inform parents/guardians of associated gynecologic anomalies?

Clinical Question

<u>P (population/problem)</u>	Among parents/guardians of females with ARM,
<u>I (intervention)</u>	does being informed about their daughter's associated gynecologic anomalies at diagnosis of ARM
<u>C (comparison)</u>	as opposed to later in the course of her care
<u>O (outcome)</u>	increase their satisfaction with healthcare decisions related to their daughter's long-term sexual & reproductive health?

Target Population: Parents/guardians of females diagnosed with an anorectal malformation

Recommendation(s): (See Table of Recommendation Strength following references)

1. It is recommended that each patient and parent(s)/guardian(s) be offered information about all known aspects of the condition at the time of diagnosis. (*Greenburg, et al., 1984[4a]; Krahn, et al., 1993[4b]; Byrnes, et al., 2003[4a]; Aite, et al., 2006[4a]; Levetown, 2008[5a]; local consensus [5]*).
Note: One parent is quoted, "Knowing what you're dealing with is so much easier, even if you don't want to know." (*Krahn, 1993[5]*).
2. Due to insufficient specific evidence regarding satisfaction with long term healthcare decisions, it is recommended that research be conducted in our target population to evaluate the impact of offering information about all known aspects of the condition at the time of initial diagnosis of ARM.

Discussion/summary of evidence:

The literature search identified 20 articles, 8 of which were determined to have some relevance to the PICO question. Several make general recommendations for the informing interview, including aspects such as the content, timing, and setting (*Aite, et al., 2006[4a]; Byrnes, et al., 2003[4a]; Girgis, 1995[5a]; Krahn, et al., 1993[4b]; Greenberg, et al., 1984[4a]; Levetown, 2008[5a]; Starke, et al., 2002[4b]*).

Only one of the reviewed studies involved patients diagnosed with ARM (*Aminoff, et al., 2006[4b]*), but it did not specifically discuss the timing of the informing interview about associated gynecologic anomalies in female patients and the resultant effect on parent satisfaction. In their descriptive study, Aminoff, et al. (2006 [4b]), report that 55% of those responding to their mailed questionnaire indicated that while they were given an explanation of their child's anorectal malformation prior to surgery, they did not feel as if information about functional prognosis for later in life was given in the same detail, if given at all. Fifty seven percent of respondents stated that they were not informed about Bowel Management. Functional gynecologic outcomes were not addressed in the survey. The authors also report that parents describe positive outcomes as a result of working with a multidisciplinary health care team later in life and would have preferred that this approach had been emphasized to them at the time of diagnosis. Starke et al. (2002 [4b]), report that

parents of girls with Turner Syndrome would like specific details of the diagnosis to be explained by experts in the field, supporting the results of the Aminoff work in another population.

Four descriptive studies, (Greenburg, et al., 1984[4a]; Krahn, et al., 1993[4b]; Byrnes, et al., 2003[4a]; Aite, et al., 2006[4a]), and one expert opinion article, (Levetown, 2008[5a]), discuss the timing of the disclosure of a medical or surgical diagnosis and universally recommend that information be shared with parents as soon as it is available.

The collective body of reviewed evidence (Aite, et al., 2006[4a]; Aminoff, et al., 2006[4b]; Byrnes, et al., 2003[4a]; Girgis, 1995[5a]; Krahn, et al., 1993[4b]; Greenberg, et al, 1984[4a]; Levetown, 2008[5a]; Starke, et al., 2002[4b]) emphasizes that disclosure of the details of a medical condition be individualized. “Breaking Bad News: Consensus Guidelines for Medical Practitioners” (Girgis, 1995[5a]), outlines the pros and cons of three main categories of disclosure: non-disclosure, full disclosure, and individualized disclosure. It concludes that individualized disclosure is the preferred method of sharing information.

Although we were unable to find any published articles addressing the disclosure of details of congenital gynecologic anomalies, we believe that the data derived from research done in areas of other congenital anomalies and chronic conditions can be applied to our patient population.

Grade for the body of evidence related to PICO: low (due to no studies available that answer the clinical question or directly involve the patient population)

Health Benefits:

- Promotes trust between the health care provider and family (Girgis, 1995[5a]; Krahn, et al., 1993[4b])
- Enhances the ability of the parent/ guardian to make informed treatment decisions(Girgis, 1995[5a]; Greenberg, et al, 1984[4a])
- Earlier diagnosis/detection of gynecologic complications thereby reducing risk to reproductive organs and fertility (local consensus)

Risks:

- Potential for the inability of the parent/guardian to process large amounts of information(Girgis, 1995[5a])
- Possible short term negative emotional impact on parents/guardians(Girgis, 1995[5a]; Starke, et al., 2002[4b])

References/citations: (evidence grade in []; see *Table of Evidence Levels following references*)

Aite, L., Zaccara, A., Trucchi, A., Nahom, A., Iacobelli, B., & Bagolan, P. (2006). Parents' informational needs at the birth of a baby with a surgically correctable anomaly. *Pediatric Surgery International*, 22(3), 267-270. [4a]

Aminoff D., La Sala E., Zaccara A., AIMAR (Italian Parent's Association of Children Born with ARM). (2006). Follow-up of anorectal anomalies: The Italian parents' and patients' perspective. *Journal of Pediatric Surgery*, 41(4), 837-841. [4b]

Byrnes, A., Berk, N., Cooper, M., & Marazita, M. (2003). Parental evaluation of informing interviews for cleft lip and/or palate. *Pediatrics*, 112(2; 2), 308. [4a]

Girgis A. & Sanson-Fisher, S. (1995). Breaking bad news: Consensus guidelines for medical practitioners. *J Clin Oncol*, 13(9), 2449-2456. [5a]

Krahn, G. L., Hallum, A., & Kime, C. (1993). Are there good ways to give `Bad news`? *Pediatrics*, 91(3), 578-582. [4b]

Greenberg, L.W., Jewett, L.S., Gluck R.S., Champion, L.A., Leikin, S. L., Altieri, M.F., Lipnick, R.N. (1984). Giving information for a life-threatening diagnosis. *AJDC*, (138), 649-653. [4a]

Levetown, M., & American Academy of Pediatrics Committee on Bioethics. (2008). Communicating with children and families: From everyday interactions to skill in conveying distressing information. *Pediatrics*, 121(5), e1441-60. [5a]

Local Consensus, During BESt development time frame. [5]

Starke, M., Wikland, K. A., & Moller, A. (2002). Parents' experiences of receiving the diagnosis of turner syndrome: An explorative and retrospective study. *Patient Education & Counseling*, 47(4), 347-354.[4b]

Note: Full tables of evidence grading system available in separate document:

- [Table of Evidence Levels of Individual Studies by Domain, Study Design, & Quality](#) (abbreviated table below)
- [Grading a Body of Evidence to Answer a Clinical Question](#)
- [Judging the Strength of a Recommendation](#) (abbreviated table below)

Table of Evidence Levels (see note above)

<i>Quality level</i>	<i>Definition</i>
1a† or 1b†	Systematic review, meta-analysis, or meta-synthesis of multiple studies
2a or 2b	Best study design for domain
3a or 3b	Fair study design for domain
4a or 4b	Weak study design for domain
5	Other: General review, expert opinion, case report, consensus report, or guideline

†a = good quality study; b = lesser quality study

Table of Recommendation Strength (see note above)

<i>Strength</i>	<i>Definition</i>
“Strongly recommended”	There is consensus that benefits clearly outweigh risks and burdens (or visa-versa for negative recommendations).
“Recommended”	There is consensus that benefits are closely balanced with risks and burdens.
No recommendation made	There is lack of consensus to direct development of a recommendation.

Dimensions: In determining the strength of a recommendation, the development group makes a considered judgment in a consensus process that incorporates critically appraised evidence, clinical experience, and other dimensions as listed below.

1. Grade of the Body of Evidence (see note above)
2. Safety / Harm
3. Health benefit to patient (*direct benefit*)
4. Burden to patient of adherence to recommendation (*cost, hassle, discomfort, pain, motivation, ability to adhere, time*)
5. Cost-effectiveness to healthcare system (*balance of cost / savings of resources, staff time, and supplies based on published studies or onsite analysis*)
6. Directness (*the extent to which the body of evidence directly answers the clinical question [population/problem, intervention, comparison, outcome]*)
7. Impact on morbidity/mortality or quality of life

Supporting information

Introductory/background information: The Colorectal Center for Children at Cincinnati Children's Hospital Medical Center provides multidisciplinary medical and surgical management for patients with anorectal malformations. Patients come to our center at different stages in their treatment. Many parents of female patients with an ARM report they have never been given the opportunity to have a detailed discussion with a health care provider about their daughter's possible associated gynecologic anomalies. Often parents are eager to have this conversation and state they wish they had been given more information earlier in the course of their daughter's treatment. We sought to discover if evidence existed to support the premise that parents would feel more comfortable making treatment decisions if they were aware of their daughter's reproductive anatomy and the possible implications this could have on her future gynecologic health.

Group/team members:

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Search strategy

1. DATABASES

OVID MEDLINE

CINAHL

OVID EBM Reviews (Cochrane) PubMed Clinical Queries

other (specify)

PsychInfo

2. SEARCH TERMS

Ano-rectal malformation, anal rectal malformation, anorectal malformation, anus-abnormalities, rectum-abnormalities, gynecologic malformations, gynecologic anomalies, gynecological abnormalities, gynecological anomalies, reproductive anomalies, congenital anomalies, informing interview, parental attitudes, parental preferences

3. LIMITS AND FILTERS (check all that apply)

English

Humans

Age Range (specify)

Publication Date Range (specify)

1990-present

Clinical Queries or other Clinical Filters (specify)

Other filters or limits (specify)

Copies of this Best Evidence Statement (BESt) are available online and may be distributed by any organization for the global purpose of improving child health outcomes. Website address: <http://www.cincinnatichildrens.org/svc/alpha/h/health-policy/ev-based/default.htm>
Examples of approved uses of the BESt include the following:

- copies may be provided to anyone involved in the organization's process for developing and implementing evidence based care;
- hyperlinks to the CCHMC website may be placed on the organization's website;
- the BESt may be adopted or adapted for use within the organization, provided that CCHMC receives appropriate attribution on all written or electronic documents; and
- copies may be provided to patients and the clinicians who manage their care.

Notification of CCHMC at HPCEInfo@cchmc.org for any BESt adopted, adapted, implemented or hyperlinked by the organization is appreciated.

Additionally for more information about CCHMC Best Evidence Statements and the development process, contact the Health Policy & Clinical Effectiveness office at: 513-636-2501 or HPCEInfo@chmcc.org.

[as appropriate, substitute *Center for Professional Excellence/Research and Evidence-based Practice office at CPE-EBP-Group@chmcc.org* for the contact information. If the BESt was developed by another group, the appropriate contact information ought to be substituted.]

Note

This Best Evidence Statement addresses only key points of care for the target population; it is not intended to be a comprehensive practice guideline. These recommendations result from review of literature and practices current at the time of their formulation. This Best Evidence Statement does not preclude using care modalities proven efficacious in studies published subsequent to the current revision of this document. This document is not intended to impose standards of care preventing selective variances from the recommendations to meet the specific and unique requirements of individual patients. Adherence to this Statement is voluntary. The clinician in light of the individual circumstances presented by the patient must make the ultimate judgment regarding the priority of any specific procedure.

Reviewed by Cincinnati Children's Hospital Medical Center – Evidence Federation