

ERN eUROGEN registry FAQ

1.7 Anorectal Malformations

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1. QUESTIONS

Question: Is an anteriorly displaced anus an anorectal malformation?

Answer: No.

Question: Is a fistula ani an anorectal malformation?

Answer: No.

Common Data Elements – Anorectal malformation patients

2.7.3.1.7 ITEM: **Rare or complex disease**

Question: When is it considered a VACTERL association?

Answer: When a patient has at least three or more associated Vertebral, Cardiac, Tracheoesophageal, Renal or Limb abnormalities, besides the anorectal malformation. For example, a patient with a perineal fistula, an absent coccyx, and only one kidney is considered to have VACTERL association. Rib anomalies do not fall under the VACTERL association.

If a patient has 1 or 2 associated abnormalities, it's not considered a VACTERL association, please select 'Isolated anorectal malformation'.

Question: What is meant with 'isolated' anorectal malformation?

Answer: This means that the anorectal malformation is not part of an association or syndrome. This does not mean that the patient cannot have any other abnormalities. For example, a patient with a rectovestibular fistula also has a tracheoesophageal fistula. This patient should still be registered under isolated anorectal malformation.

2.7.3.1.7.2 ITEM: **Type of anorectal malformation**

Question: What type is a 'bucket handle'?

Answer: You can register this as a perineal fistula.

Patient characteristics

6.4.1 ITEM: **What was the patient referred for?**

Question: What is the difference between the patient referred for surgery, second opinion, and the patient referred for transfer of care?

Answer: If the patient is transferred for surgery, then only the reconstructive surgery will be done in your HCP and afterwards the patient will go back to their own hospital for follow-up. If the patient is referred for second opinion, then no surgery takes place in your HCP and the doctors at your HCP have only given advice during a consultation. If the patient is transferred for care, then all care, including all follow-up, will be done in your HCP.

Diagnostics and associated abnormalities

6.10.3 ITEM: **Were any abnormalities found on this (first) X-ray of the spine and/or sacrum?**

Question: DO abnormalities affecting the coccyx, such as an absent coccyx, fall under sacral abnormalities?

Answer: Yes, you can register this under Sacral malformations, and in this case > partial sacral agenesis.

6.5.3 ITEM: **What abnormalities were found on this first ultrasound of the kidneys and bladder?**

Question: Are 'echo-dense' kidneys considered an abnormality?

Answer: No.

Question: Is a dilated collecting/pelvicalyceal system or dilated pyelum an abnormality?

Answer: Yes, you can register this as hydronephrosis.

Question: Is a small kidney an abnormality?

Answer: Yes, you can register this as a dysplastic/hypoplastic kidney.

6.13.3.1 ITEM: **What abnormalities were found on the first echocardiogram?**

Question: Is a bicuspid aortic valve an abnormality?

Answer: Yes, you can register this under 'Other' under ITEM 6.13.3.1.1, or ITEM 6.14.2 > Cardiac abnormalities > Other, depending if this was found on the first echocardiogram (6.13.3.1.1) or later (6.14.2)

Question: What if an abnormality was found that was not listed in ITEMS 6.5 to 6.13, where do I register this?

Answer: You can register it under ITEM 6.14 "Have any other abnormalities (not previously mentioned) been found?".

Surgery and treatments

6.15 ITEM: **Does or did the patient have a stoma / enterostomy?**

Question: The patient's stoma was constructed in a different hospital, is this okay?

Answer: This is okay. If you know that the patient has had a stoma, but you do not know the date, type, and bowel section, you can enter 01-01-1900 for date and select "I don't know / no information in medical file" for the other two.

6.15.2 ITEM: **What type of stoma / enterostomy was constructed?**

Question: Is a double barrel stoma the same as a split/divided stoma?

Answer: Yes.

6.17 ITEM: **Has the patient undergone reconstructive surgery?**

Question: The patient has undergone reconstructive surgery in another hospital, is this okay?

Answer: No. Only patients that have undergone reconstructive surgery in your HCP can be included in the registry.

6.17.2 ITEM: **What type of reconstructive surgery has the patient undergone?**

Question: The patient has undergone a modified PSARP with preservation of the external sphincter, how do I register this?

Answer: You can still register this under PSARP.

Question: Is a TERPT a type of reconstructive surgery?

Answer: Yes, but usually for Hirschsprung disease, however it may be possible that the patient has both ARM and Hirschsprung disease, so you should register it under 'Other'.

6.17.4 ITEM: **Were there any complications after the surgery?**

Question: The patient had some issues after surgery, such as episodes of hypoglycaemia (low blood sugar), difficulties getting off the oxygen, episodes of bradycardia (low heart rate), they resolved. Should they be registered as complications?

Answer: No. These complications are related to the anaesthesia required for the surgery, but not directly related to the surgical procedure itself. Especially if they resolved or did not require any interventions, they should not be registered. If in doubt, select "Yes" and you will see the possible options to select from to give you an idea of common complications related to the surgery. If you are still in doubt, you can always register the complication under "Other".

6.19 ITEM: **Did the patient undergo anal dilatations?**

Question: The patient underwent calibrations, is this the same?

Answer: Yes, this is the same.

6.20 ITEM: **Is the patient treated with any stool regulators?**

Question: The patient used to be treated with stool regulators, but now is not anymore, what should I select?

Answer: You can select No. You should base your selection on the information in the latest medical note.

Question: Are klysmas/enemas the same as rectal water irrigation systems?

Answer: No.

Functional outcomes

6.24 and 6.25 ITEMS: **Is the patient developing / growing well?**

Question: How old may the information regarding growth and development be?

Answer: It is okay if the information is a bit older, you may trust the information in the latest medical note that mentions growth and/or development. If you think the information is too old or untrustworthy, select "I don't know / no information in medical file".



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