ERN eUROGEN registry
Report on Expertise Area 1.7

INTRODUCTION

This report entails an analysis of the data on Expertise Area 1.7 Anorectal malformations in the ERN eUROGEN registry. This report aims to give insight in the current clinical practices using the Clinical Practice Snapshot data about the patients entered. Only HCPs that entered more than 5 patients for 1.7 Anorectal malformations were included individually in these analyses. HCPs that entered less than 5 patients, were grouped in an 'Other' HCP group. This 'Other' HCP group consists of DK02 (N=1), IT11 (N=4), IT34 (N=3) and SE01 (N=4). As patient numbers are not similar across HCPs, the results cannot be equally compared between HCPs, but the analyses give an indication of trends.

The Clinical Practice Snapshots should only contain data about the first year of treatment which starts from the date the patient first visits the ERN eUROGEN HCP for the anorectal malformation. However, sometimes information outside the 1-year window was added, and at other times, the dates are unknown. If this occurs, we interpreted this variable for this patient as 'Not performed'. An example: A patient had the first visit to the hospital (start treatment) at 23-01-2021, and the reconstructive surgery took place at 08-02-2022 (more than a year after the start of treatment). This surgery should not have been entered in the Clinical Practice Snapshot of the ERN eUROGEN registry, and we interpreted it as 'No reconstructive surgery within the first year'.

Please keep in mind that these reports are meant to inform you about some general treatment characteristics using the Clinical Practice Snapshot data, not to perform in-depth statistical analysis. If you have any suggestions about information to add to these reports, or to delete because the information is not relevant, please let us know and it will be considered for the next report.

EA 1.7; ANORECTAL MALFORMATIONS

Descriptive statistics

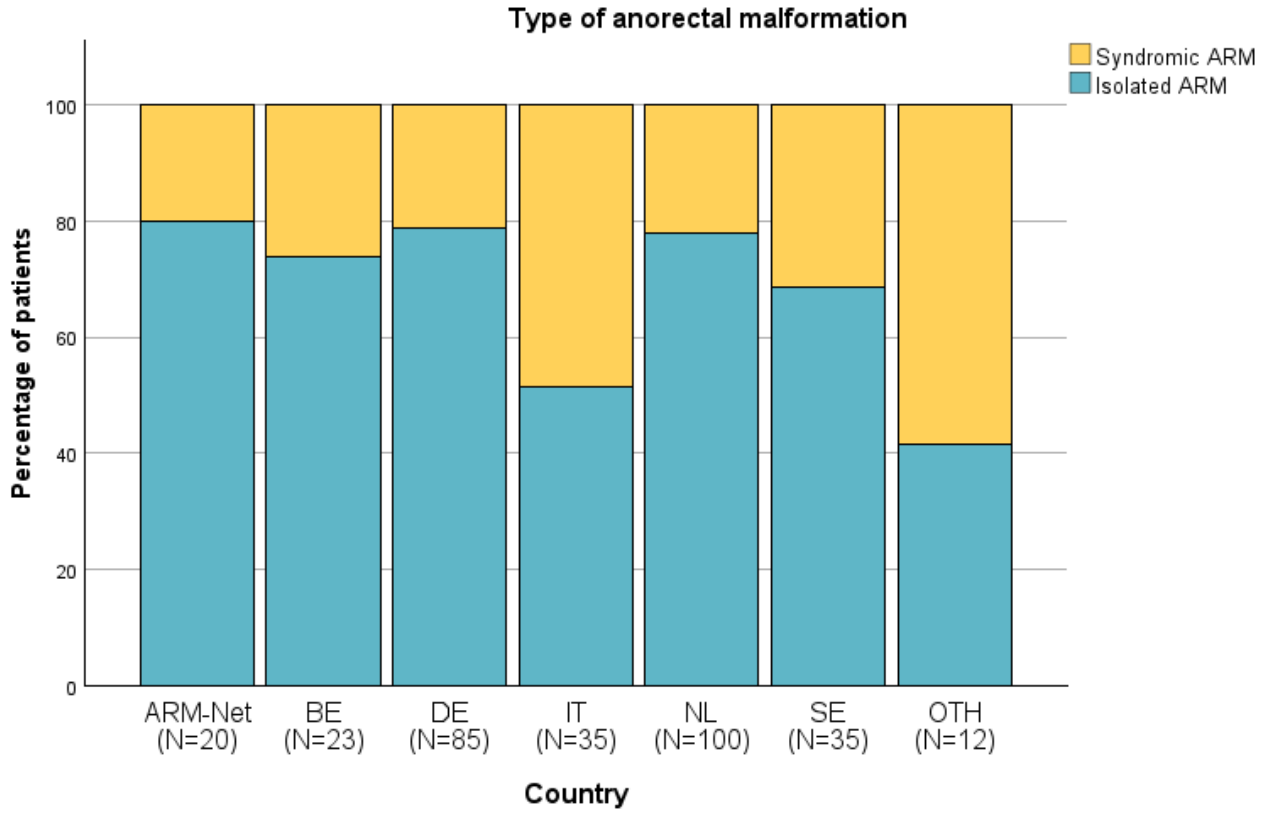
The table below provides an overview of the descriptive statistics for patients from Expertise Area 1.7 Anorectal malformations. Corresponding figures were made of the variables, and they are displayed on the next pages.

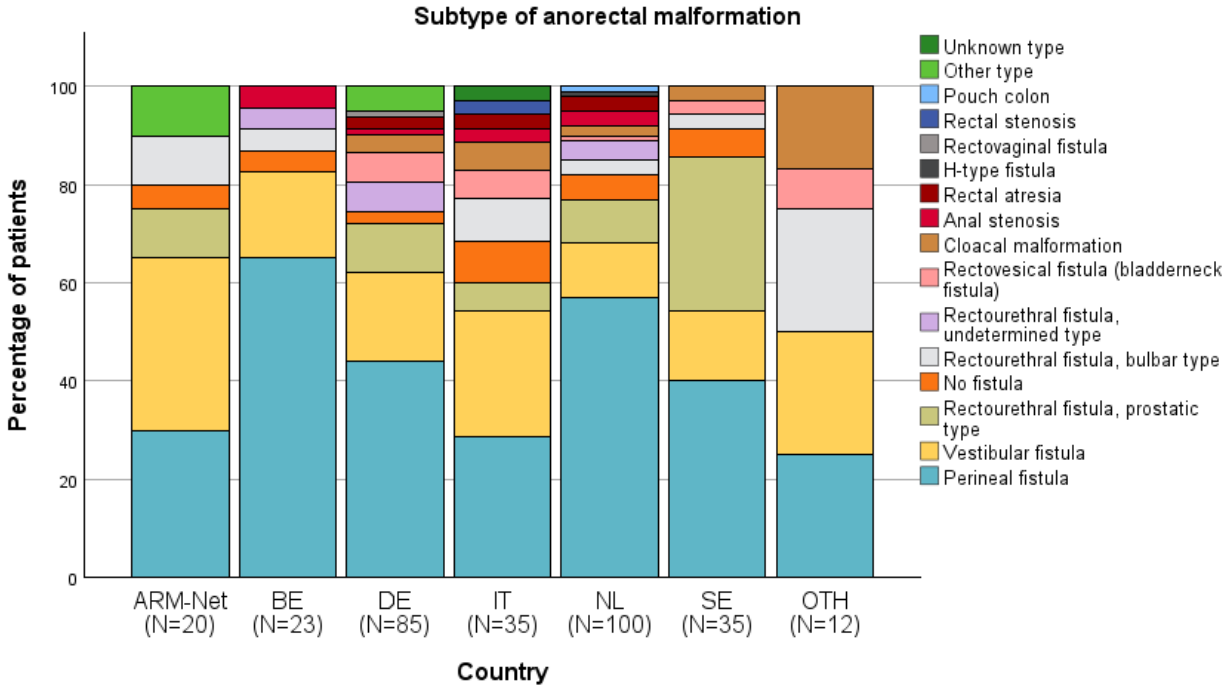
	Total	ARM-Net	Belgium	Germany		Italy		Netherlands			Sweden	Others
	N=310	ARM-Net (N=20)	BE10 (N=23)	DE09 (N=52)	DE13 (N=33)	IT05 (N=15)	IT58 (N=20)	NL01 (N=40)	NL03 (N=21)	NL09 (N=39)	SE03 (N=35)	OTH (N=12)
Type of malformation												
Non-syndromic anorectal malformation; N (%)	552 (72.6%)	16 (80.0%)	17 (73.9%)	46 (88.5%)	21 (63.6%)	6 (40.0%)	11 (55.0%)	33 (82.5%)	17 (81.0%)	28 (71.8%)	24 (68.6%)	5 (41.7%)
Syndromic anorectal malformation; N (%)	85 (27.4%)	4 (20.0%)	6 (26.1%)	6 (11.5%)	12 (36.4%)	9 (60.0%)	9 (45.0%)	7 (17.5%)*	4 (19.0%)	11 (2829%)	11 (31.4%)	7 (58.3%)
Diagnostics, timing after first visit & abnormalities												
Ultrasound of kidney and bladder												
Performed; N (%)	243 (78.4%)	16 (80.0%)	19 (82.6%)	41 (78.8%)	21 (63.3%)	12 (80.0%)	17 (85.0%)	35 (87.5%)	20 (95.2%)	21 (53.8%)	31 (88.6%)	10 (83.3%)
Days first visit to USS; Median (range)	2 days (0;323)	1 day (0;21)	0 days (0;51)	0 days (0;112)	2 days (0;274)	3 days (0;203)	0 days (0;1)	2 days (0;29)	2 days (0;57)	2 days (0;70)	5 days (0;154)	2 days (1;323)
Abnormalities found on USK : N (%)	64 (26.3%)	4 (25.0%)	5 (26.3%)	12 (29.3%)	9 (42.9%)	4 (33.3%)	5 (29.4%)	8 (22.9%)	2 (10.0%)	5 (23.8%)	4 (12.9%)	6 (60.0%)
Voiding cystourethrogram (VCUG)												
Performed; N (%)	91 (29.4%)	4 (20.0%)	13 (56.5%)	11 (21.2%)	3 (9.1%)	4 (26.7%)	4 (20.0%)	7 (17.5%)	-	25 (64.1%)	16 (45.7%)	4 (33.3%)
Days first visit to VCUG; Median (range)	41 days (0;334)	52 days (1;119)	45 days (0;263)	29 days (12;245)	8 days (2;48)	44 days (20;133)	32 days (14;175)	41 days (7;85)	-	70 days (2;158)	26 days (3;68)	22.5 days (6;334)
VUR diagnosed; N (%)	28 (30.8%)	3 (75.0%)	3 (23.1%)	6 (54.5%)	2 (66.7%)	1 (25.0%)	-	1 (14.3%)	-	5 (20.0%)	6 (37.5%)	1 (25.0%)
X-ray of spine/sacrum												
Performed; N (%)	163 (52.6%)	12 (60.0%)	17 (73.9%)	4 (7.7%)	5 (15.2%)	7 (46.7%)	18 (90.0%)	36 (90.0%)	19 (90.5%)	18 (46.2%)	18 (51.4%)	9 (75.0%)
Days first visit to X-ray; Median (range)	3 days (0;319)	0 days (0;309)	108 days (0;319)	1 day (0;1)	10 days (0;254)	1 day (0;230)	4 days (0;115)	3 days (0;71)	3 days (0;142)	2 days (0;70)	4 days (0;15)	4 days (1;46)
Abnormalities found on X-ray: N (%)	49 (30.1%)	4 (33.3%)	8 (47.1%)	4 (100%)	3 (60.0%)	2 (28.6%)	6 (33.3%)	9 (25.0%)	1 (5.3%)	4 (22.2%)	5 (27.8%)	3 (33.3%)
Ultrasound of spine/sacrum												
Performed; N (%)	205 (66.1%)	11 (55.0%)	13 (56.5%)	34 (65.4%)	18 (54.5%)	4 (26.7%)	16 (80.0%)	32 (80.0%)	20 (95.2%)	19 (48.7%)	29 (82.9%)	9 (75.0%)
Days first visit to USS&S; Median (range)	3 days (0;263)	1 day (0;162)	5 days (0;263)	4 days (0;217)	6 days (0;55)	12 days (141)	9 days (0;142)	2 days (0;29)	3 days (0;57)	2 days (0;70)	5 days (0;49)	3 days (1;40)
Abnormalities found on USS&S; N (%)	57 (27.8%)	4 (36.4%)	2 (15.4%)	10 (29.4%)	10 (55.6%)	1 (25.0%)	5 (31.3%)	9 (28.1%)	1 (5.0%)	4 (21.1%)	6 (20.7%)	5 (55.6%)
MRI of spine/sacrum												
Performed; N (%)	72 (23.2%)	9 (45.0%)	4 (17.4%)	7 (13.5%)	14 (42.4%)	3 (20.0%)	4 (20.0%)	12 (30.0%)	3 (14.3%)	2 (5.1%)	10 (28.6%)	4 (33.3%)
Days first visit to MRI; Median (range)	71 days (1;340)	82 days (6;294)	70 days (17;311)	74 days (3;249)	97 days (1;340)	187 days (148;240)	30 days (10;212)	22 days (7;98)	67 days (65;71)	2 days (1;2)	104 days (9;292)	103 days (12;313)
Abnormalities found on MRI; N (%)	47 (65.3%)	6 (66.7%)	2 (50.0%)	7 (100%)	10 (71.4%)	3 (100%)	3 (75.0%)	8 (66.7%)	1 (33.3%)	1 (50%)	5 (50.0%)	1 (25.0%)
Echocardiogram												
Performed; N (%)	220 (71.0%)	17 (85.0%)	20 (87.0%)	22 (42.3%)	16 (48.5%)	12 (80.0%)	20 (100%)	35 (87.5%)	20 (95.2%)	18 (46.2%)	31 (88.6%)	9 (75.0%)
Days first visit to ECG; Median (range)	1 day (0;285)	1 (0;285)	1 day (0;108)	5 days (0;102)	1 day (0;71)	3 days (0;147)	1 day (0;116)	2 days (0;180)	10 days (0;102)	1 day (0;70)	0 days (0;57)	1 day (0;5)
Abnormalities found on ECG; N (%)	98 (44.5%)	10 (58.8%)	3 (15.0%)	10 (45.5%)	12 (75.0%)	4 (33.3%)	14 (70.0%)	20 (57.1%)	1 (5.0%)	14 (77.8%)	7 (22.6%)	3 (33.3%)
Surgery and treatment												
Reconstructive surgery												
Performed within 1 year after 1st visit; N (%)	265 (85.5%)	17 (85.0%)	18 (78.3%)	40 (76.9%)	25 (75.8%)	15 (100%)	17 (85.5%)	33 (82.5%)	21 (100%)	36 (92.3%)	32 (91.4%)	11 (91.7%)
Performed in own HCP; N (%)	238 (89.8%)	15 (88.2%)	17 (94.4%)	34 (85.0%)	18 (72.0%)	12 (80.0%)	17 (100%)	32 (97.0%)	17 (81.0%)	35 (97.1%)	31 (97.2%)	10 (90.9%)
Age at surgery; Median (range)	103 days (0;6042)	141 days (0;378)	175 days (1;414)	109 days (1;6042)	36 days (1;751)	189 days (56;314)	71 days (3;154)	123 days (45;1713)	32 days (1;216)	109 days (1;253)	42 days (1;920)	70 days (2;339)
Stoma/enterostomy												
Patients with stoma/enterostomy; N (%)	95 (30.6%)	6 (30.0%)	7 (30.4%)	11 (21.2%)	8 (24.2%)	3 (20.0%)	7 (35.0%)	10 (25.0%)	8 (38.1%)	7 (17.9%)	21 (60.0%)	7 (58.3%)
Patient with closed stoma; N (%)	80 (84.2%)	4 (66.7%)	6 (85.7%)	9 (81.8%)	6 (75.0%)	3 (100%)	5 (71.4%)	10 (100%)	8 (100%)	7 (100%)	16 (90.5%)	6 (85.7%)
Days stoma was in place; Median (range)	167 days (32;343)	236 days (94;240)	140 (68;317)	218 days (69;303)	195 days (51;209)	256 days (151;343)	184 days (111;224)	154 days (32;265)	206 days (167;299)	139 days (69;337)	120 days (78;225)	147 days (124;339)
Stool regulators												
Treated with stool regulators; N (%)	193 (63.5%)	8 (42.1%)	17 (17.3%)	34 (65.4%)	22 (66.7%)	9 (60.0%)	12 (60.0%)	33 (82.5%)	11 (52.4%)	18 (46.2%)	23 (69.7%)	6 (66.7%)

Type of malformation

Type of anorectal malformation

Most patients were diagnosed with an isolated form of anorectal malformation. Most patients were diagnosed with a perineal fistula, followed by vestibular fistula.

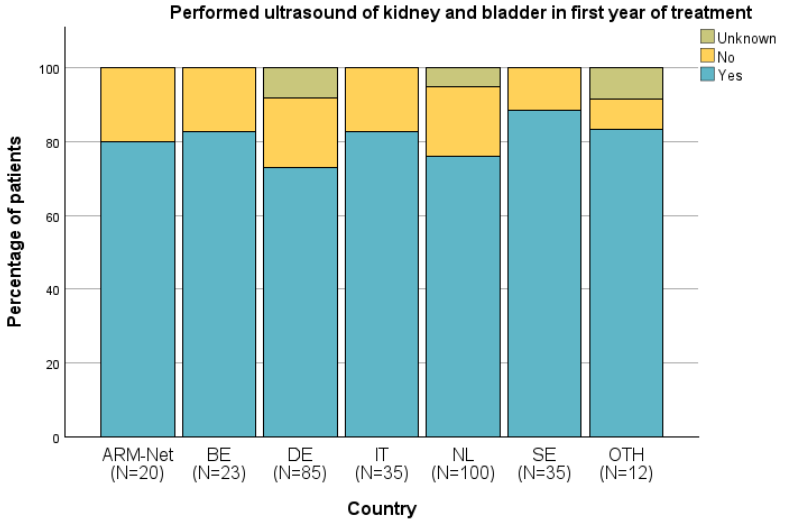


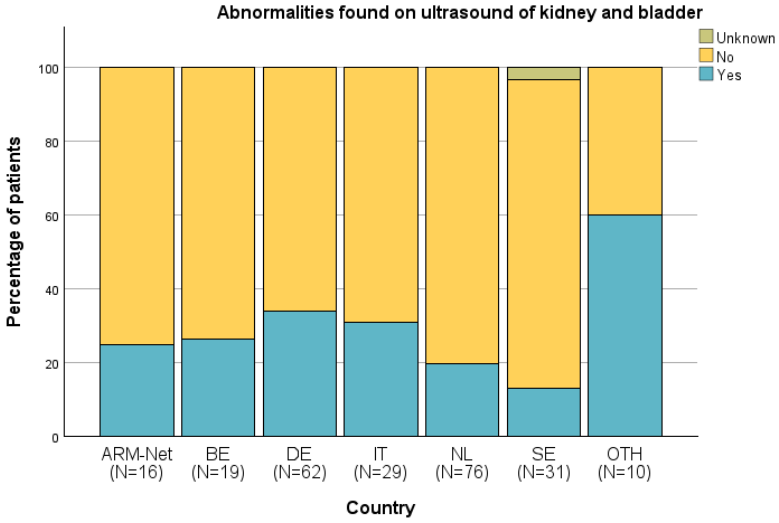


Diagnostics, timing after first visit & abnormalities

Ultrasounds of kidney and bladder and abnormalities found in the first year of treatment

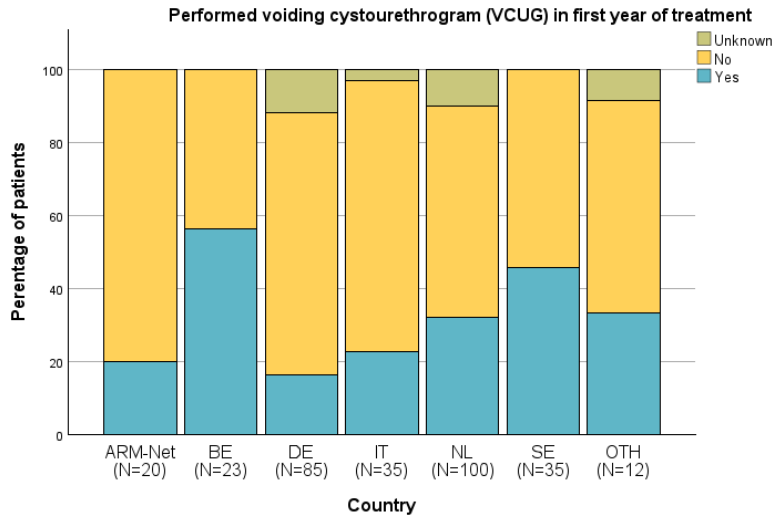
An ultrasound of the kidney and bladder was performed in most patients. When an ultrasound was performed, no abnormalities were found in most patients.

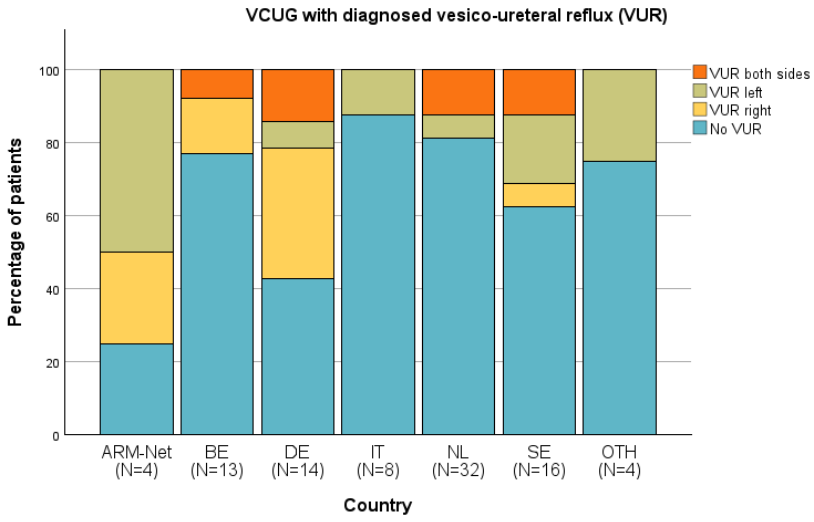




Voiding cystourethrogram (VCUG) and vesicoureteral reflux (VUR) in the first year of treatment

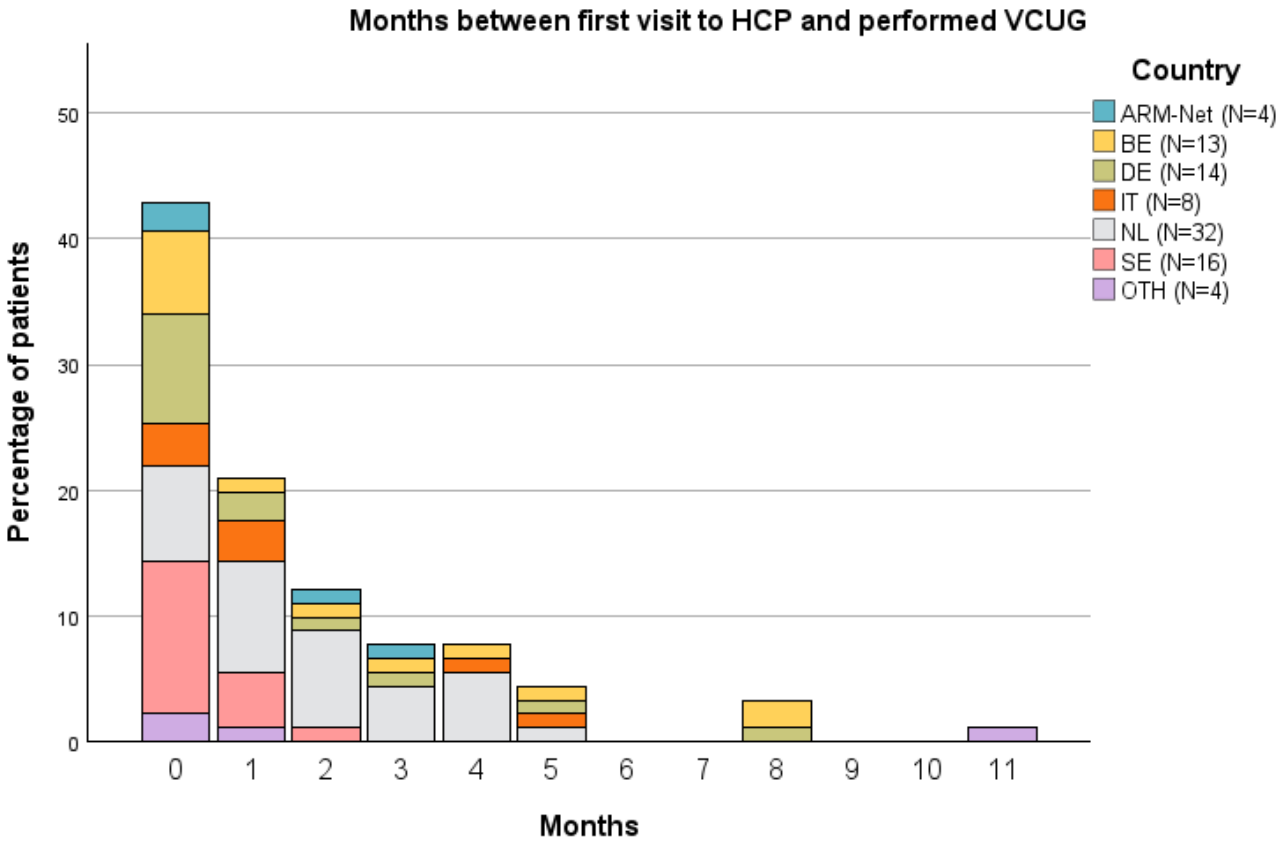
VCUG was not performed in most patients. If a VCUG was performed, no VUR was diagnosed in most cases.





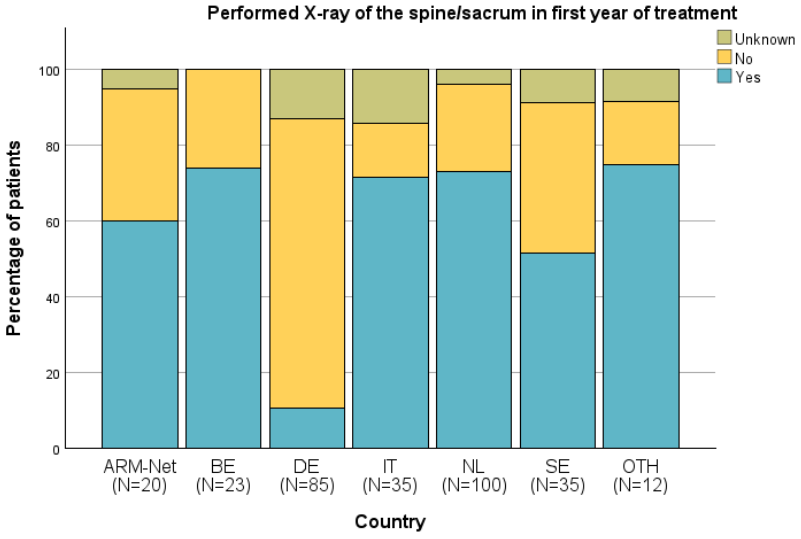
Months between first visit to HCP and VCUG

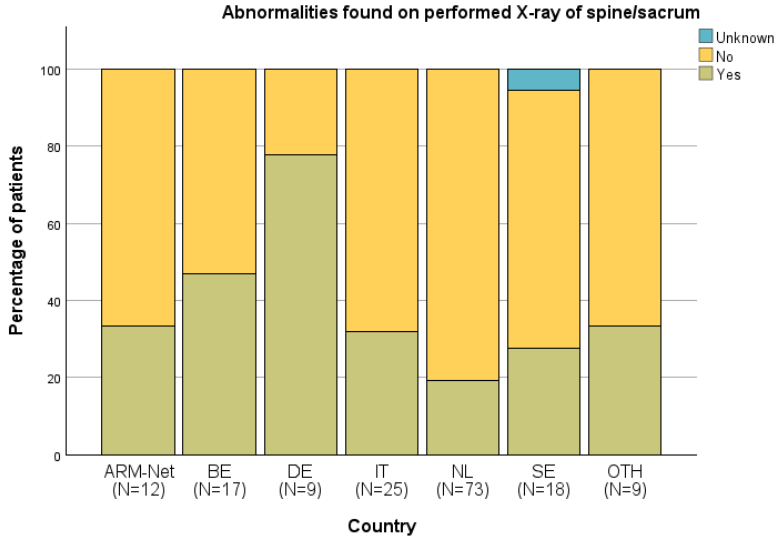
Almost all VCUGs were performed within the first half year after the first visit to the HCP.



X-ray of spine/sacrum and abnormalities found in the first year of treatment

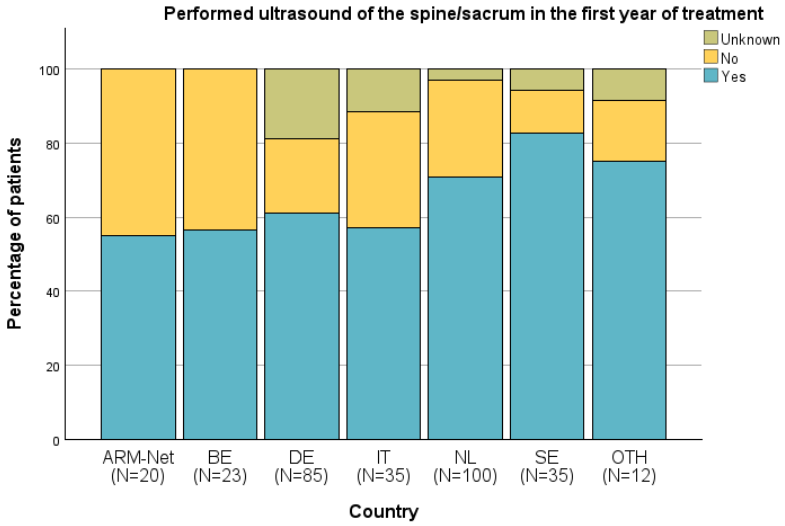
An X-ray was performed in part of the patients. Most X-rays did not reveal any abnormalities.

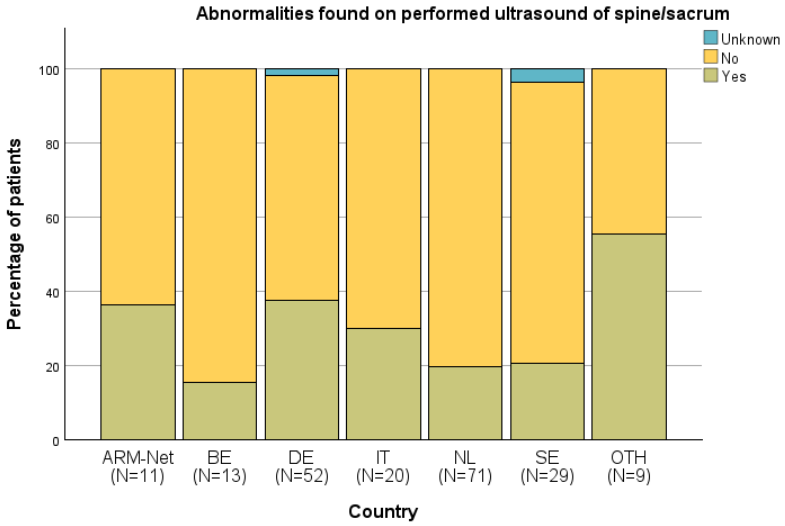




Ultrasound of spine/sacrum and abnormalities found in the first year of treatment

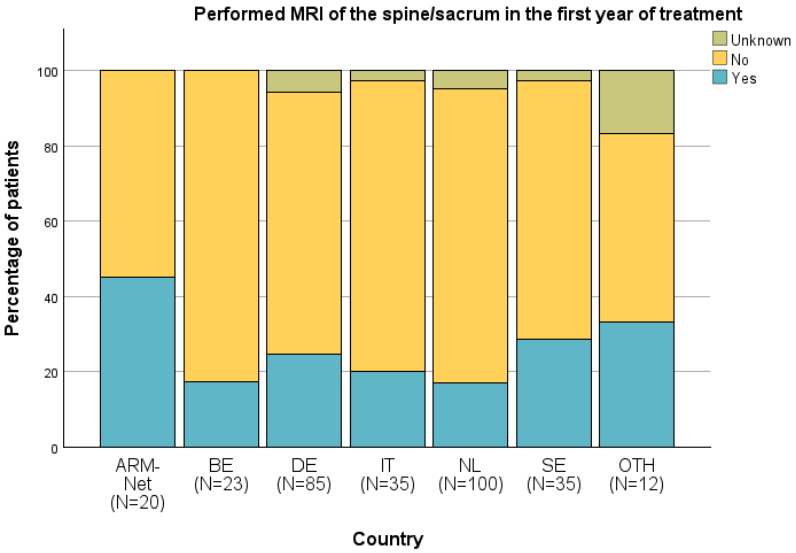
In a majority patients, an ultrasound of the spine/sacrum was performed. Most of the time, no abnormalities were found.

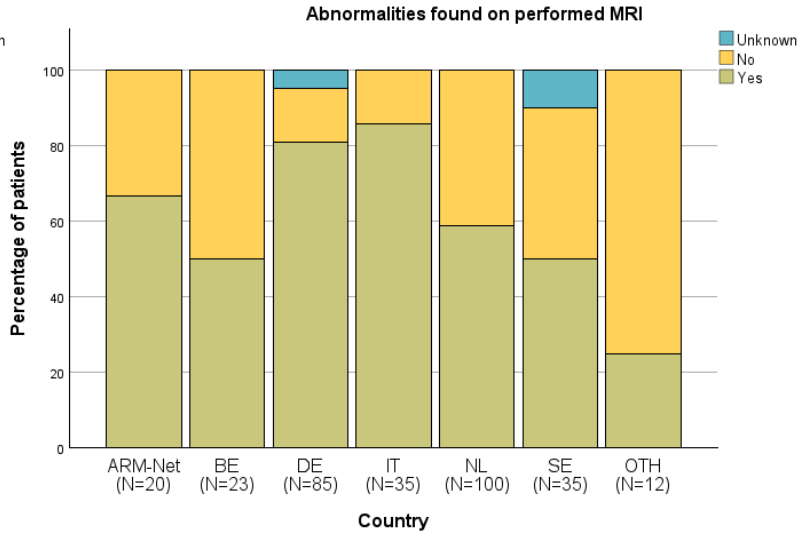




MRI of spine/sacrum and abnormalities found in the first year of treatment

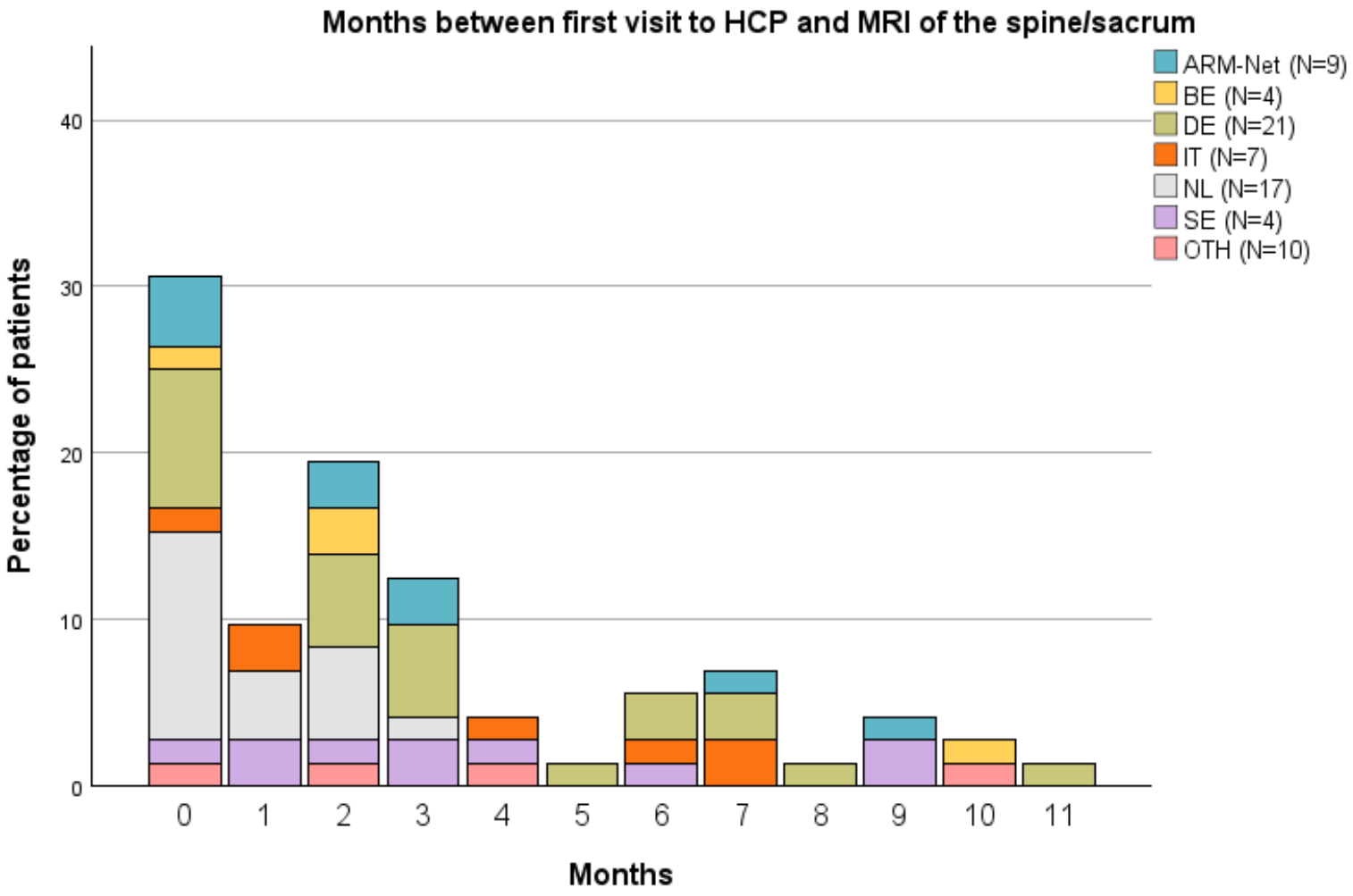
For most patients, no MRI was performed. If an MRI was done, it did reveal an abnormality in most cases.





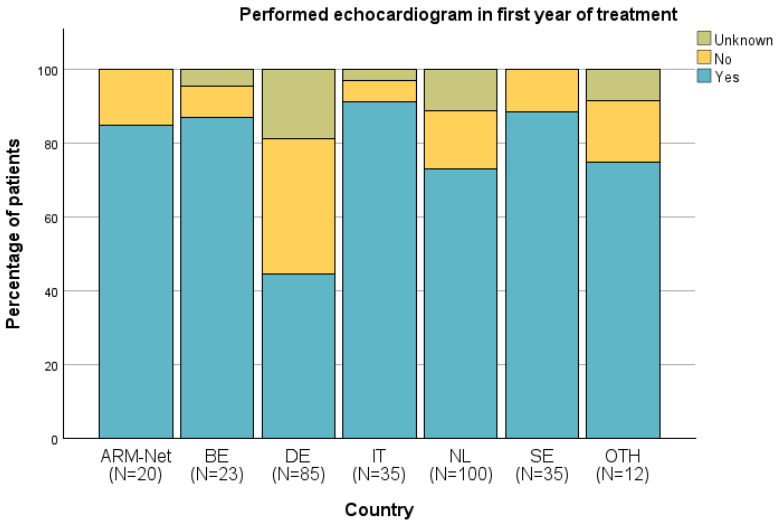
Months between first visit to HCP and MRI of spine/sacrum

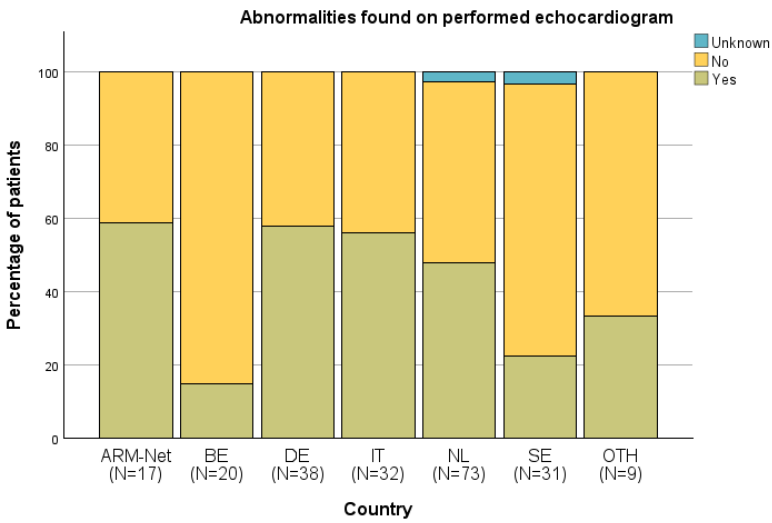
The MRIs that were performed, were mostly performed within the first half year after the first visit to the HCP.



Echocardiogram (ECG) of spine/sacrum and abnormalities found in the first year of treatment

For most patients, an ECG was performed. In part of the patients, abnormalities were found.

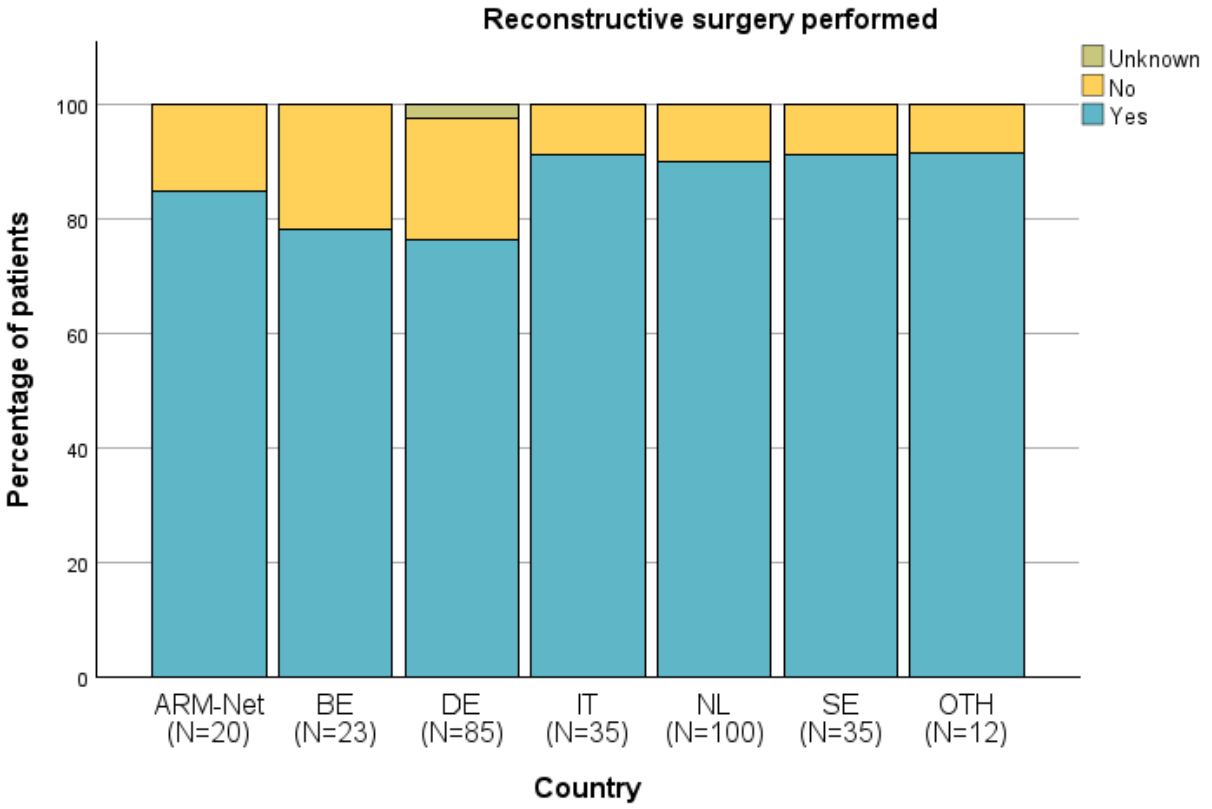




Surgery and treatment

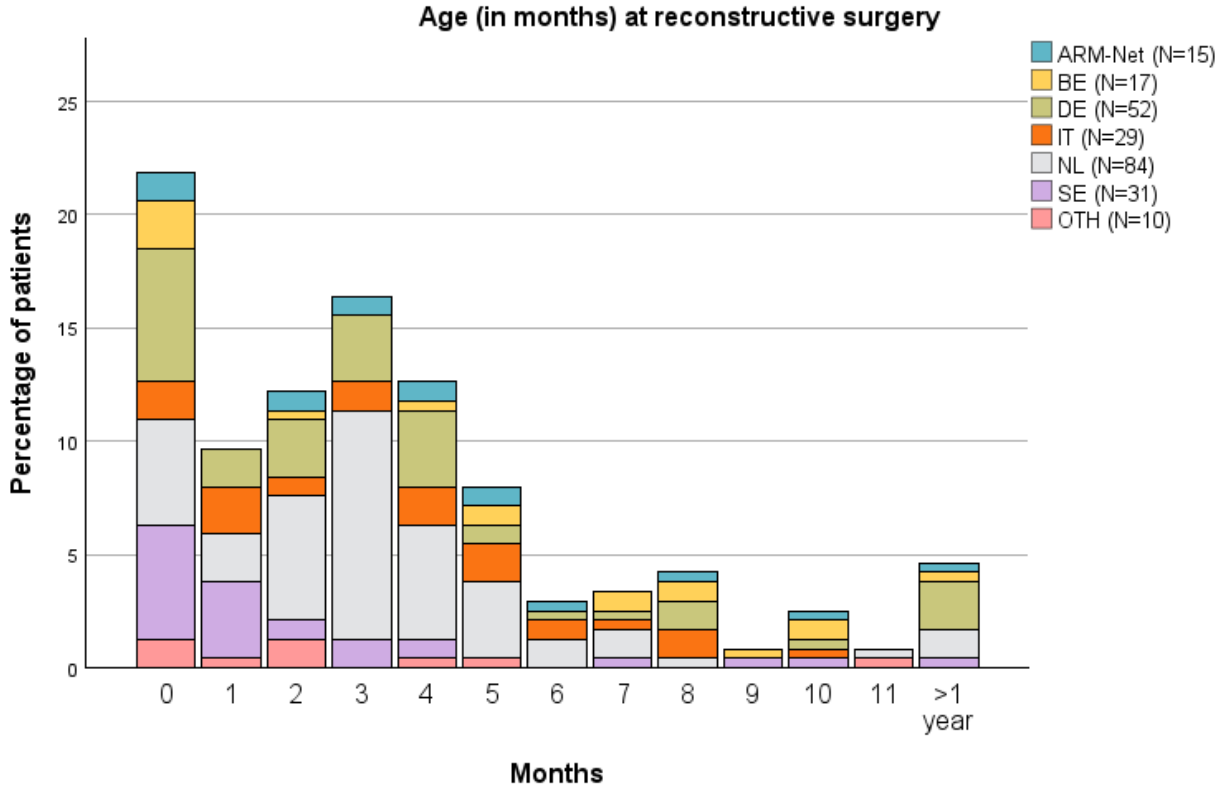
Reconstructive surgery in the first year of treatment

Most patients had a reconstructive surgery for their anorectal malformation within one year after their 1st visit to the HCP.



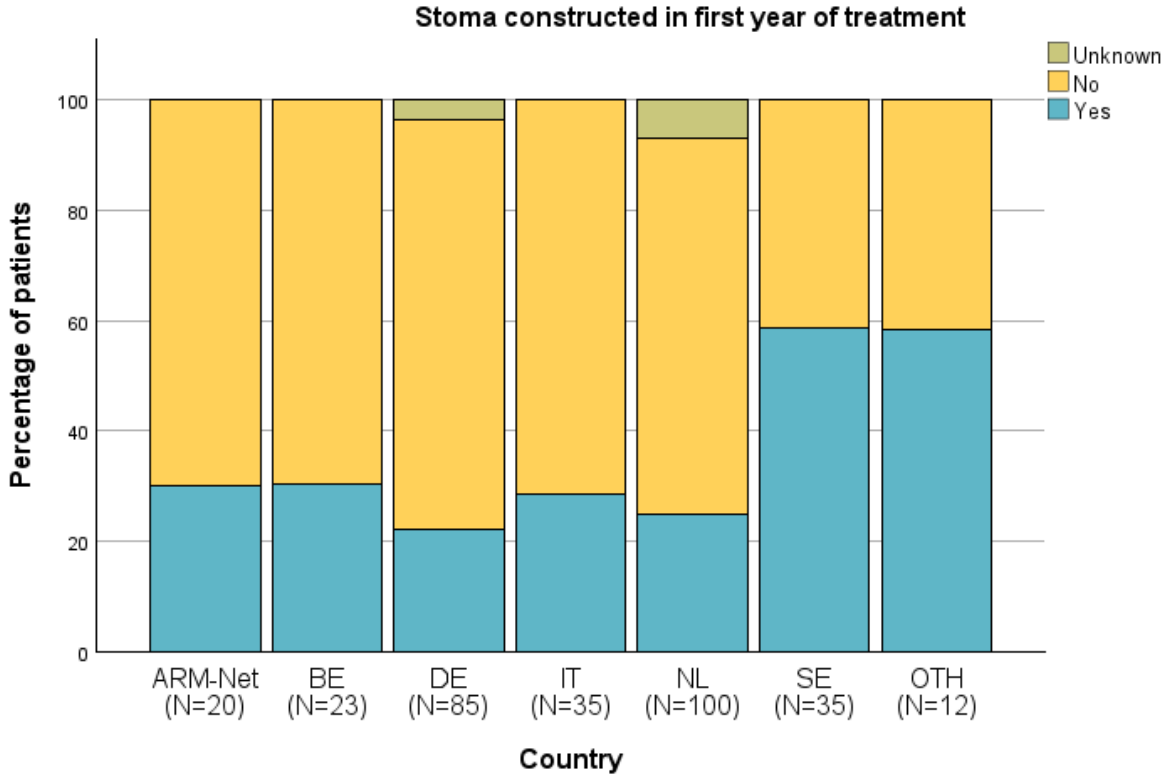
Age at reconstructive surgery when performed in the first year of treatment in own HCP

Most patients had the reconstructive surgery within the first half year of their life.



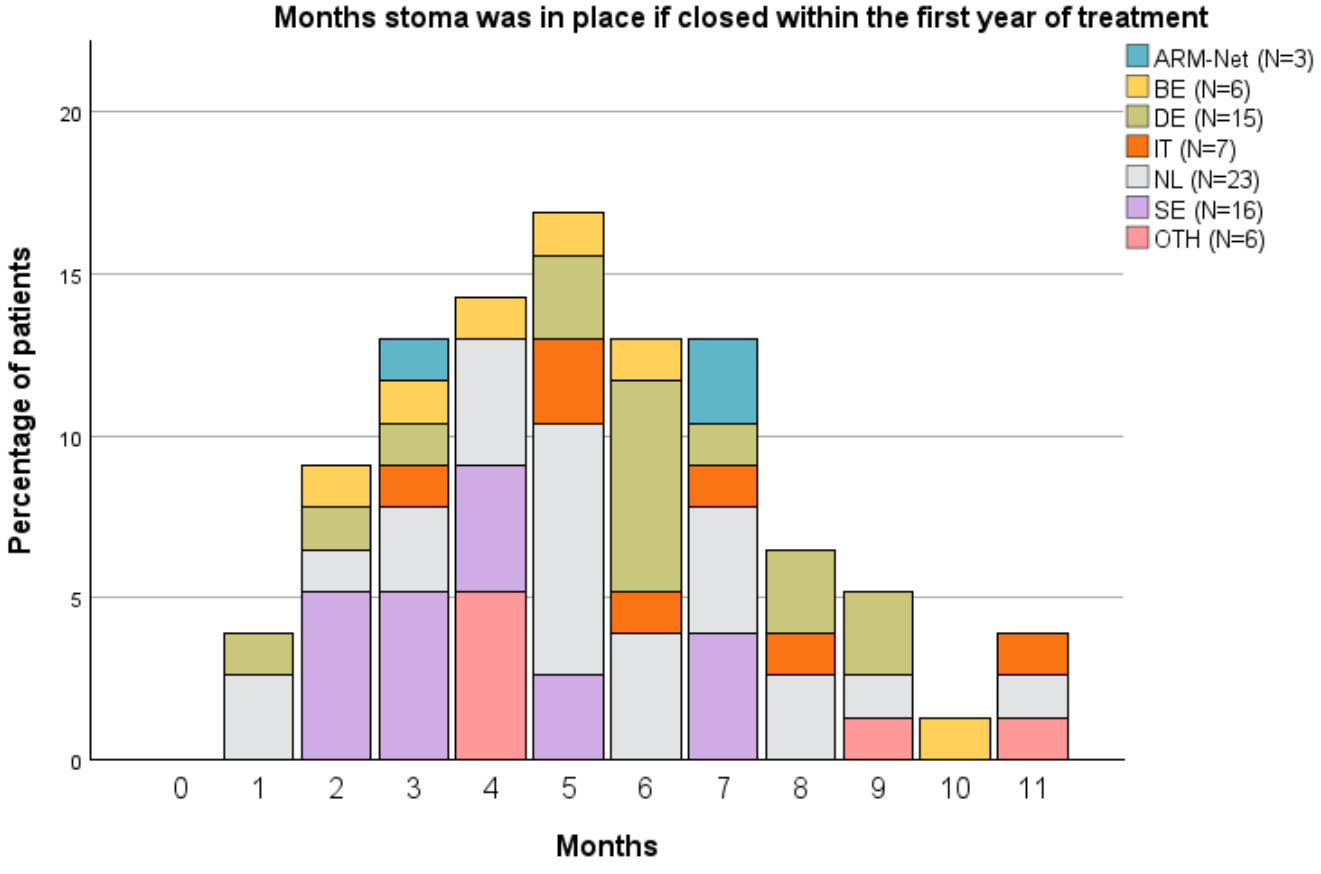
Stoma construction in the first year of treatment

A minority of patients received a stoma during their first year of treatment.



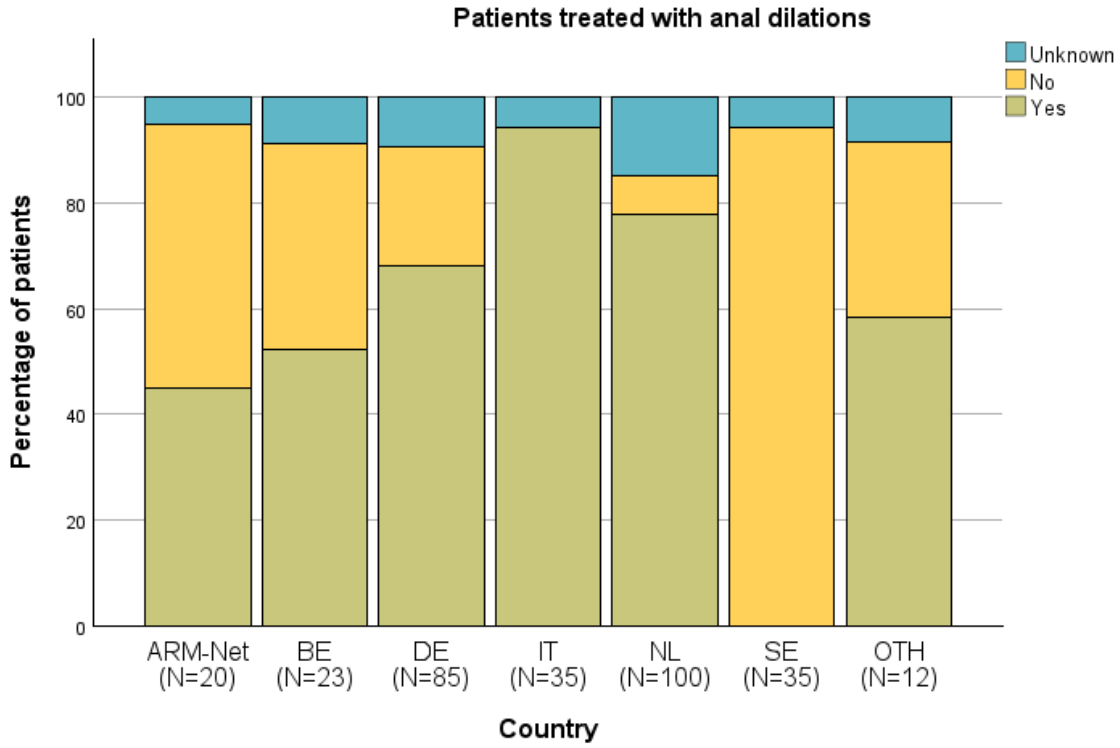
Months stoma was in place if already closed

There is a large variety in how long the stoma was in place for patients with a closed stoma, but most stoma's were in place for three to seven months.



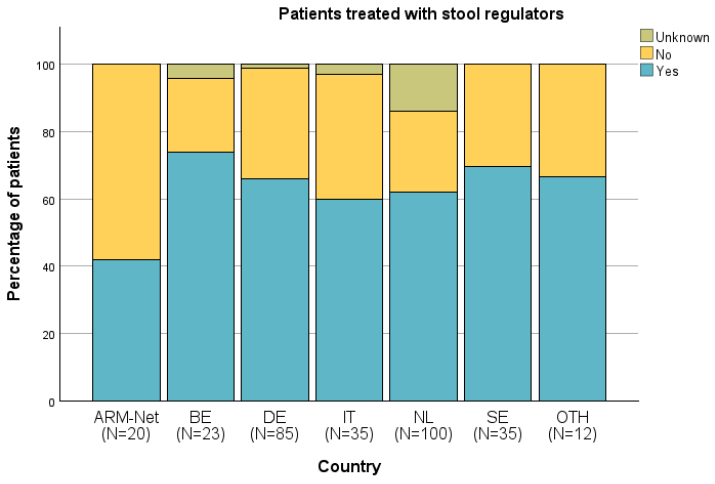
Anal dilations

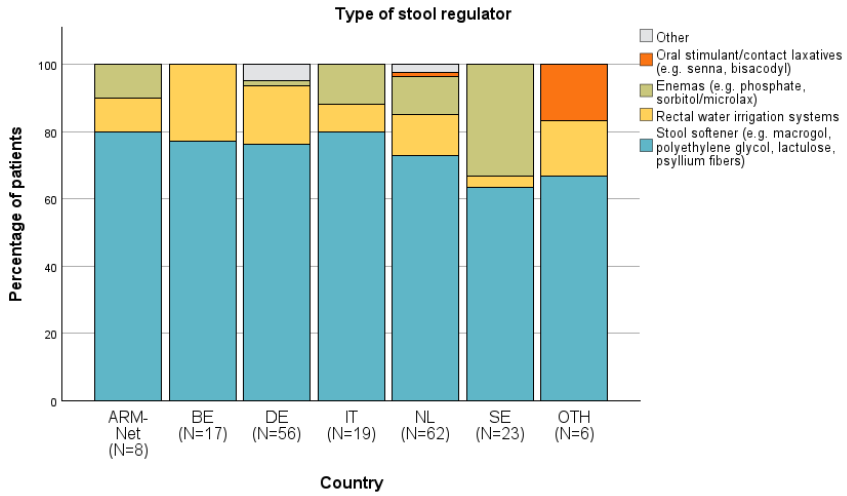
A majority of the patients received anal dilations.



Treatment with stool regulators

Most patients were treated with stool regulators. If treated with a stool regulator, stool softener was the most common used regulator.





http://eurogen-ern.eu/

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