ERN eUROGEN registry
Report on Expertise Area 1.2

INTRODUCTION

This report entails the 2025 ERN eUROGEN registry analysis of the Expertise Area 1.2 Classic Bladder Exstrophy. 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 1.2 Classic Bladder Exstrophy were included in the analyses. HCPs that entered less than 5 patients, were grouped in an 'Other' HCP. This 'Other' HCP consists of DE22 (N=1) and ES17 (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 two years of treatment which starts from the date the patient first visits the ERN eUROGEN HCP for the classic bladder exstrophy. However, sometimes information outside the 2-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 an ultrasound of the kidney took place at 08-02-2023 (more than two years after the start of treatment). This ultrasound should not have been entered in the Clinical Practice Snapshot of the ERN eUROGEN registry. If this information was there, we interpreted it as 'No ultrasound performed during the first two years of treatment'.

Please keep in mind 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 1.2; CLASSIC BLADDER EXSTROPHY

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 N=44	Czechia CZ02 (N=8)	Germany		Italy IT34 (N=5)	The Netherlands NL09 (N=6)	Sweden SE01 (N=5)	Others OTH (N=5)
			DE19 (N=9)	DE36 (N=6)				
General characteristics								
Birth sex								
Male; N (%)	28 (63.6%)	7 (87.5%)	7 (77.8%)	4 (66.7%)	3 (60.0%)	2 (33.3%)	2 (40.0%)	3 (60.0%)
Female; N (%)	16 (36.4%)	1 (12.5%)	2 (22.2%)	2 (33.3%)	2 (40.0%)	4 (66.7%)	3 (60.0%)	2 (40.0%)
Primary diagnosis and treatment								
Initial wound coverage								
No coverage; N (%)	1 (2.33%)	-	-	-	1 (20.0%)	-	-	-
Diaper; N (%)	12 (28.6%)	-	-	6 (100%)	-	4 (66.7%)	2 (40.0%)	-
Wet gauze; N (%)	23 (54.8%)	2 (25.0%)	9 (100%)	6 (100%)	3 (60.0%)	-	2 (40.0%)	1 (33.3%)
Other; N (%)	10 (23.8%)	-	-	6 (100%)	-	1 (16.7%)	3 (60.0%)	-
Unknown; N (%)	11 (26.2%)	6 (75.0%)	-	-	2 (40.0%)	1 (16.7%)	-	2 (66.7%)
								-
Initial diagnostics								
Creatinine measurement child; N (%)	16 (36.4%)	1 (12.5%)	1 (11.1%)	5 (83.3%)	4 (80.0%)	3 (50.0%)	1 (20.0%)	1 (20.0%)
Culture bladder plate; N (%)	3 (6.8%)	-	-	-	-	3 (50.0%)	-	-
Ultrasound of kidneys; N (%)	34 (77.3%)	4 (50.0%)	9 (100%)	6 (100%)	4 (80.0%)	4 (66.7%)	5 (100%)	2 (40.0%)
X-ray of pelvis; N (%)	14 (31.8%)	-	-	-	1 (20.0%)	5 (83.3%)	5 (100%)	3 (60.0%)
Other diagnostics/imaging; N (%)	11 (25.0%)	-	6 (66.7%)	-	3 (60.0%)	-	2 (40.0%)	-
Unknown; N (%)	7 (15.9%)	4 (50.0%)	-	-	1 (20.0%)	-	-	2 (40.0%)
Primary closure surgery								
Primary closure surgery performed; N (%)	40 (90.0%)	6 (75.0%)	9 (100%)	6 (100%)	5 (100.0%)	6 (100%)	5 (100%)	3 (60.0%)
Procedures at primary closure surgery								
Primary closure only; N (%)	18 (45.0%)	6 (100%)	2 (22.2%)	5(83.3%)	3 (60.0%)	-	2 (40.0%)	-
Primary closure with osteotomy; N (%)	11 (27.5%)	-	-	-	1 (20.0%)	6 (100%)	3 (60.0%)	1 (33.3%)
Bladder neck reconstruction; N (%)	3 (7.5%)	-	-	-	3 (60.0%)	-	-	-
Epispadias reconstruction; N (%)	23 (57.5%)	1 (16.7%)	7 (77.8%)	6 (100%)	1 (20.0%)	2 (33.3%)	4 (80.0%)	2 (66.7%)
Genital mobilization in female; N (%)	3 (7.5%)	-	2 (22.2%)	-	1 (20.0%)	-	-	-
Inguinal hernia repair; N (%)	14 (35.0%)	-	7 (77.8%)	5 (83.3%)	-	1 (16.7%)	1 (20.0%)	-
Introitus plasty; N (%)	1 (2.5%)	-	-	-	1 (20.0%)	-	-	-
Soft in-tissue mobilization; N (%)	1 (2.5%)	-	-	-	1 (20.0%)	-	-	-
Symphysis approximation without osteotomy; N (%)	25 (62.5%)	6 (100%)	9 (100%)	6 (100%)	2 (40.0%)	-	2 (40.0%)	-
Umbilicus reconstruction; N (%)	23 (57.5%)	-	9 (100%)	6 (100%)	-	3 (50.0%)	5 (100%)	-
Ureteral reimplant; N (%)	2 (5.0 %)	-	-	-	1 (20.0%)	1 (16.7%)	-	-
Other primary surgical procedure; N (%)	2 (5.0%)	-	-	1 (16.7%)	1 (20.0%)	-	-	-
Unknown; N (%)	1 (2.5%)	-	-	-	1 (20.0%)	-	-	-

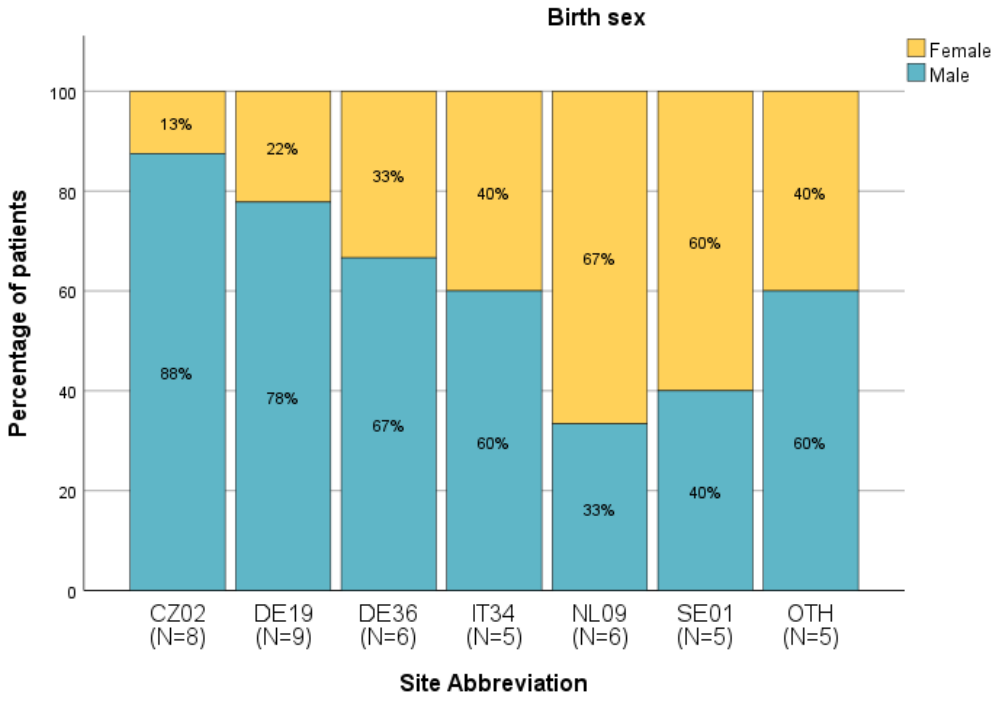
	Total N=44	Czechia CZ02 (N=8)	Germany		Italy IT34 (N=5)	The Netherlands NL09 (N=6)	Sweden SE01 (N=5)	Others OTH (N=5)
			DE19 (N=9)	DE36 (N=6)				
Location of primary closure surgery								
Own HCP; N (%)	33 (82.5%)	4 (66.7%)	9 (100%)	5 (83.3%)	3 (60.0%)	6 (100%)	3 (60.0%)	3 (100%)
Other HCP; N (%)	6 (15.0%)	2 (33.3%)	-	1 (16.7%)	1 (20.0%)	-	2 (40.0%)	-
Unknown; N (%)	1 (2.5%)	-	-	-	1 (20.0%)	-	-	-
Age at primary closure surgery; Median (range, days)	41 days (1;9959)	2 days (1;270)	52 days (34;57)	67 days (41;110)	3 days (1;98)	10 days (4;22)	67 days (32;99)	8 days (2;9959)
Type of immobilization								
None; N (%)	1 (2.5%)	-	-	-	-	-	-	1 (33.3%)
Mermaid; N (%)	27 (69.2%)	-	9 (100%)	6 (100%)	3 (60.0%)	5 (83.3%)	2 (40.0%)	2 (50.0%)
Bryant's traction; N (%)	8 (20.5%)	4 (66.7%)	-	-	-	1 (26.7%)	3 (60.0%)	-
Unknown; N (%)	4 (10.3%)	2 (33.3%)	-	-	2 (40.0%)	-	-	-
Post-surgical treatment								
Type of drainage								
Suprapubic catheter; N (%)	30 (75.0%)	4 (66.7%)	9 (100%)	6 (100%)	-	5 (83.3%)	5 (100%)	1 (33.3%)
1 ureteral splint; N (%)	6 (15.0%)	-	-	-	1 (20.0%)	3 (50.0%	-	2 (66.7%)
2 ureteral splints; N (%)	32 (80.0%)	4 (66.7%)	9 (100%)	6 (100%)	2 (40.0%)	5 (83.3%)	5 (100%)	1 (33.3%)
Transurethral catheter; N (%)	28 (70.0%)	3 (50.0%)	8 (88.9%)	6 (100%)	3 (60.0%)	5 (83.3%)	3 (60.0%)	-
Wound drain; N (%)	22 (55.0%)	2 (33.7%)	9 (100%)	6 (100%)	-	-	5 (100%)	-
Other type of drainage; N (%)	6 (15.0%)	-	5 (55.6%)	1 (16.7%)	-	-	-	-
Unknown; N (%)	4 (10.0%)	2 (33.3%)	-	-	2 (40.0%)	-	-	-
Post-surgical antibiotics treatment; N (%)	29 (72.5%)	4 (66.7%)	9 (100%)	6 (100%)	3 (60.0%)	1 (16.7%)	5 (100%)	5 (100%)
Type of antibiotic treatment								
Trimethoprim; N (%)	17 (60.7%)	-	8 (88.9%)	3 (50.0%)	-	1 (100%)	4 (100%)	5 (100%)
Amoxicillin clavulanic acid; N (%)	2 (7.1%)	-	1 (11.1%)	-	1 (33.3%)	-	-	-
Amoxicillin; N (%)	1 (3.6%)	-	-	1 (16.7%)	-	-	-	-
Other type; N (%)	5 (17.9%)	1 (25.0%)	-	2 (33.3%)	2 (66.7%)	-	-	-
Unknown; N (%)	3 (10.7%)	3 (75.0%)	-	-	-	-	-	-
Involved specialists								
Type of involved specialist								
Endocrinologist; N (%)	1 (2.3%)	-	-	-	-	1 (16.7%)	-	-
Nurse specialist; N (%)	22 (50.0%)	2 (25.0%)	9 (100%)	-	-	5 (83.3%)	5 (100%)	1 (20.0%)
Orthopedist; N (%)	20 (45.5%)	-	7 (77.8%)	-	2 (40.0%)	6 (100%)	5 (100%)	-
Paediatric nephrologist; N (%)	10 (22.7%)	3 (37.5%)	-	-	1 (20.0%)	6 (100%)	-	-
Paediatric psychologist; N (%)	13 (29.5%)	-	9 (100%)	2 (33.3%)	-	2 (33.3%)	-	-
Paediatric urologist/surgeon; N (%)	39 (88.6%)	5 (62.5%)	9 (100%)	6 (100%)	4 (80.0%)	6 (100%)	5 (100%)	4 (80.0%)
Social worker; N (%)	14 (31.8%)	-	8 (88.9%)	-	-	5 (83.3%)	-	1 (20.0%)
Other specialist; N (%)	6 (13.6%)	2 (25.0%)	-	-	1 (20.0%)	3 (50.0%)	-	-
Unknown; N (%)	4 (9.1%)	3 (37.5%)	-	-	1 (20.0%)	-	-	-

	Total N=44	Czechia CZ02 (N=8)	Germany		Italy IT34 (N=5)	The Netherlands NL09 (N=6)	Sweden SE01 (N=5)	Others OTH (N=5)
			DE19 (N=9)	DE36 (N=6)				
Additional diagnostics								
Additional diagnostics performed; N (%)	44 (100%)	8 (100%)	9 (100%)	6 (100%)	5 (100%)	6 (100%)	5 (100%)	5 (100%)
Micturition cystourethrography (MCG) ; N (%)	8 (18.2%)	1 (12.5%)	-	-	2 (40.0%)	4 (66.7%)	1 (20.0%)	-
Serum creatinine; N (%)	16 (36.4%)	2 (25.0%)	7 (77.8%)	-	4 (80.0%)	-	3 (60.0%)	-
Ultrasound of kidneys; N (%)	35 (79.5%)	4 (50.0%)	9 (100%)	6 (100%)	4 (80.0%)	6 (100%)	4 (80.0%)	2 (40.0%)
Urine culture; N (%)	18 (40.0%)	2 (25.0%)	4 (44.4%)	-	2 (40.0%)	5 (83.3%)	3 (60.0%)	2 (40.0%)
Urodynamic study (UDS) ; N (%)	2 (4.5%)	2 (25.0%)	-	-	-	-	-	-
Video urodynamic study (VUDS) ; N (%)	2 (4.5%)	2 (25.0%)	-	-	-	-	-	-
Other additional diagnostics; N (%)	10 (22.7%)	5 (62.5%)	-	1 (16.7%)	2 (40.0%)	1 (16.7%)	-	1 (20.0%)
Unknown; N (%)	6 (13.6%)	1 (12.5%)	-	-	1 (20.0%)	-	2 (40.0%)	2 (40.0%)
Diagnosed hydronephrosis if ultrasound performed; N (%)	7 (20.0%)	2 (50.0%)	2 (22.2%)	-	1 (25.0%)	-	1 (25.0%)	1 (50.0%)
Diagnosed VUR if VUDS/MCG performed; N (%)	7 (70.0%)	1 (66.7%)	-	-	1 (50.0%)	3 (75.0%)	1 (100%)	-
Urine cultures performed; N (%)	18 (40.9%)	2 (25.0%)	4 (44.4%)	-	2 (40.0%)	5 (83.3%)	3 (60.0%)	2 (40.0%)
0 positive cultures; N (%)	1 (5.6%)	-	1 (25.0%)	-	-	-	-	-
1 positive culture; N (%)	3 (16.7%)	-	1 (25.0%)	-	-	2 (40.0%)	-	-
2-5 positive cultures; N (%)	6 (33.3%)	1 (50.0%)	2 (50.0%)	-	1 (50.0%)	1 (20.0%)	1 (33.3%)	-
> 5 positive cultures; N (%)	4 (22.2%)	-	-	-	-	2 (40.0%)	-	2 (100%)
Unknown; N (%)	4 (22.2%)	1 (50.0%)	-	-	1 (50.0%)	-	2 (66.7%)	-

General characteristics

Birth sex

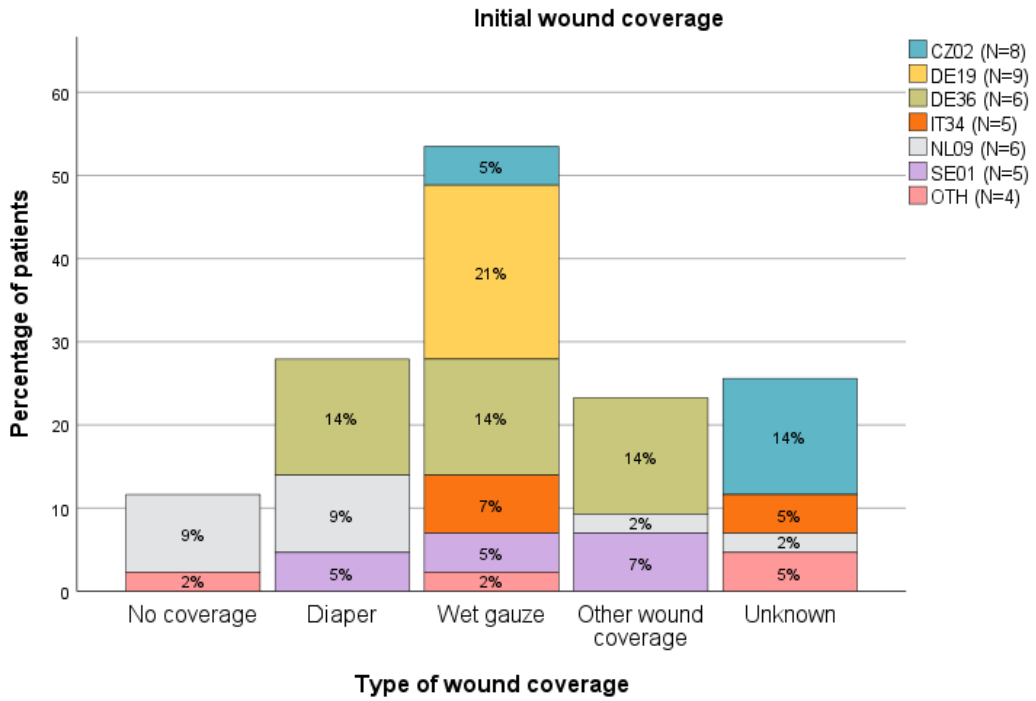
Among the registered patients, more males than females were included, although some HCPs registered more females than males.



Primary diagnosis and treatment

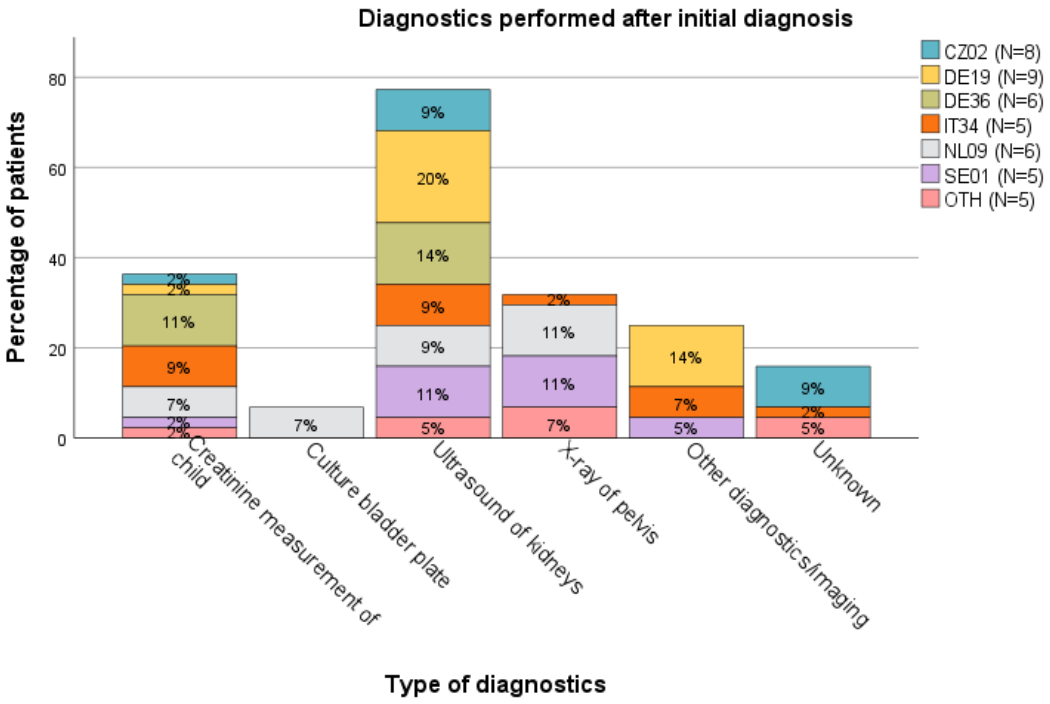
Initial wound coverage

Over half of the patients had wet gauze as initial wound coverage. Diapers and other wound coverage products were used in approximately 25% of patients. Numbers add up to more than 100% because some patients had more than one type of initial wound coverage. Some patients did not have any initial wound coverage.



Diagnostics performed after initial diagnosis

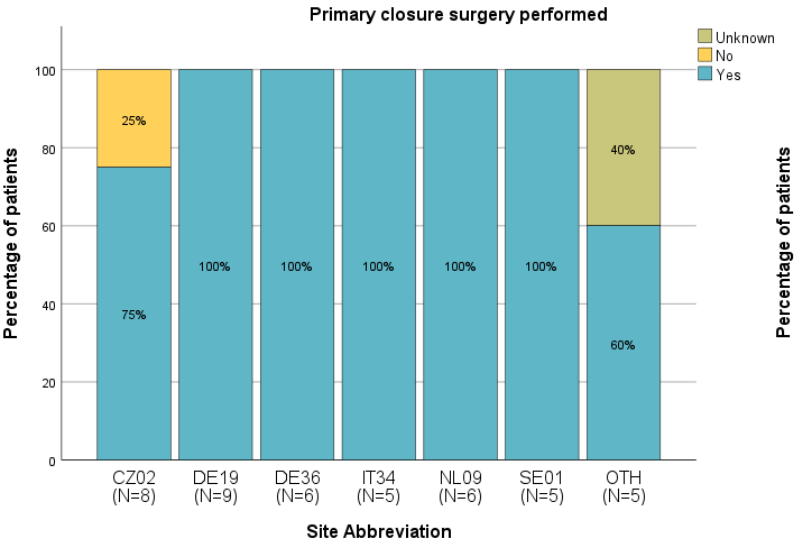
Almost all patients received an ultrasound of the kidneys during the initial diagnosis process, whereas a culture bladder plate was only performed in a small number of patients. Again, numbers add up to more than 100% because some patients had more than one type of diagnostics after the initial diagnosis.

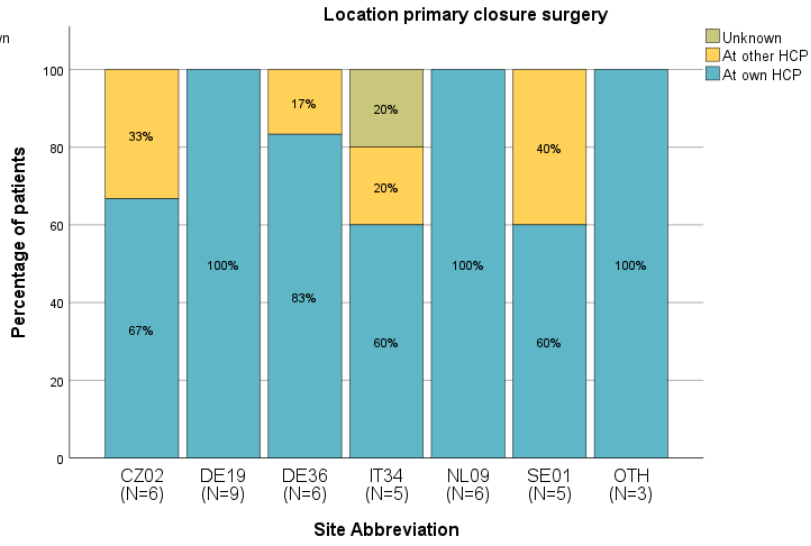


Primary closure surgery

Primary closure surgery performed and location of primary closure surgery

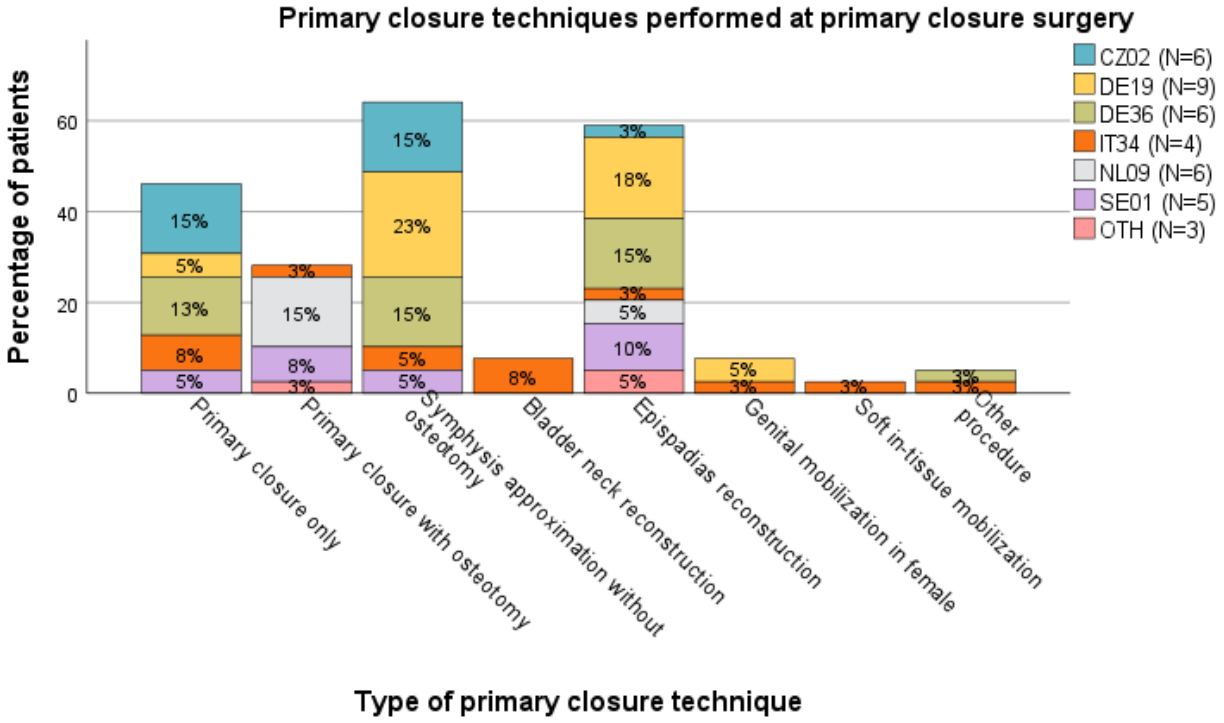
All but 4 patients had a primary closure surgery during their first two years of treatment. Most of those surgeries took place in the ERN eUROGEN HCP that registered the patient.





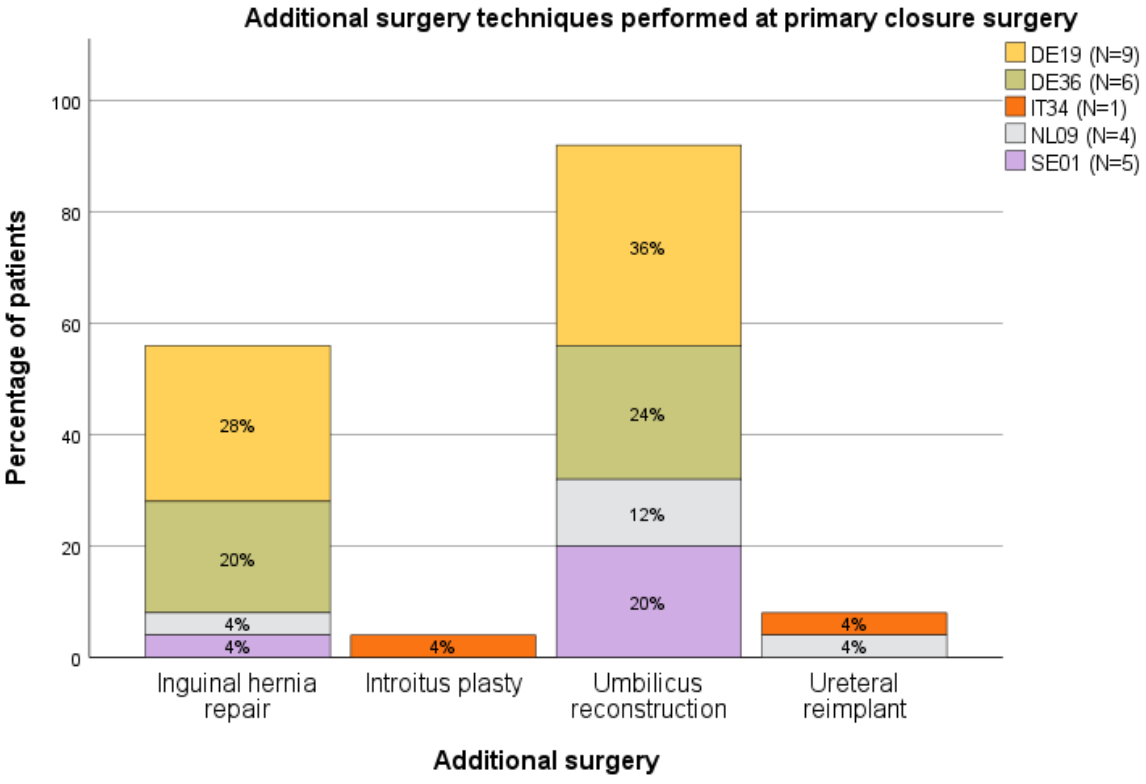
Primary closure techniques performed at primary closure surgery

At the primary closure surgery, many procedures can be performed. Regarding primary closure techniques, most common are the symphysis approximation without osteotomy and primary closure only. Soft-in tissue mobilization was only performed in a minimum of patients. Numbers add up to more than 100% because some patients had more than one type of primary closure technique at primary closure surgery.



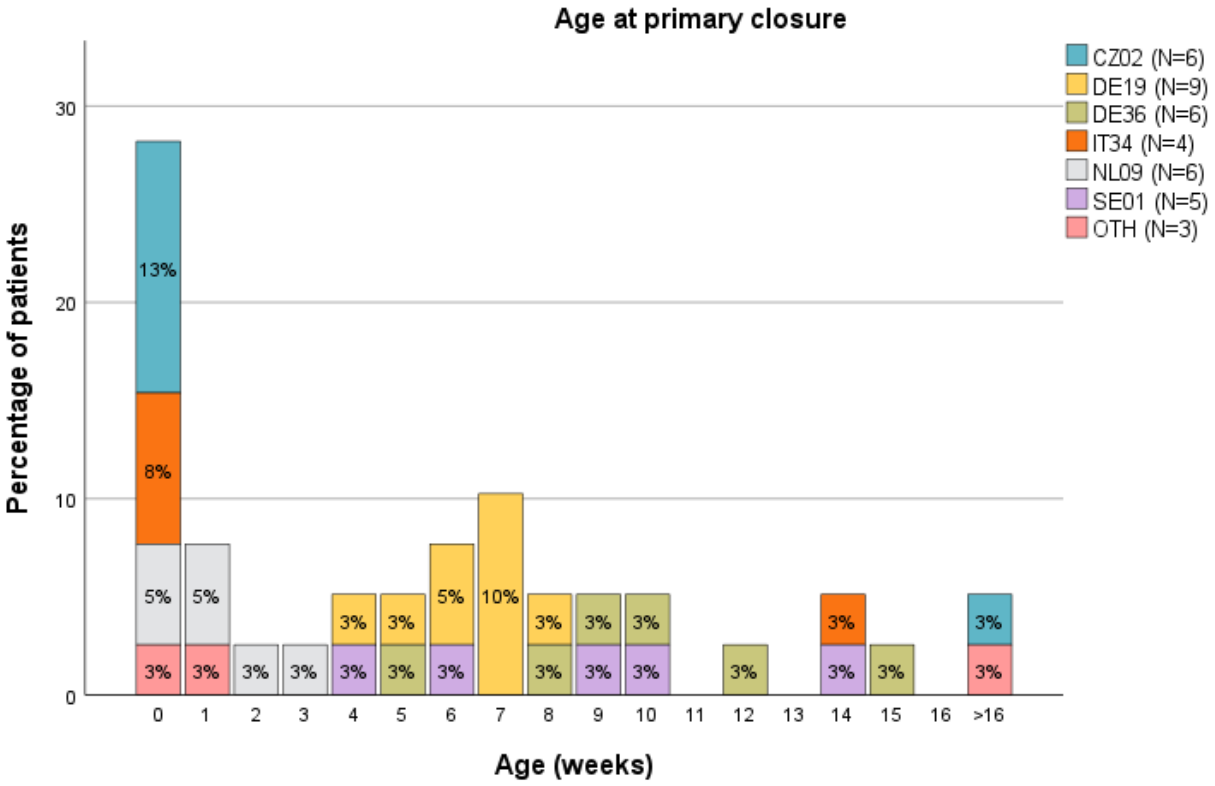
Additional surgery techniques performed at primary closure surgery

Regarding additional surgery techniques, most common are umbilicus reconstruction and inguinal hernia repair. Introitus plasty and ureteral reimplants were only performed in a minimum of patients. Numbers add up to more than 100% because some patients had more than one type of additional primary closure technique at primary closure surgery.



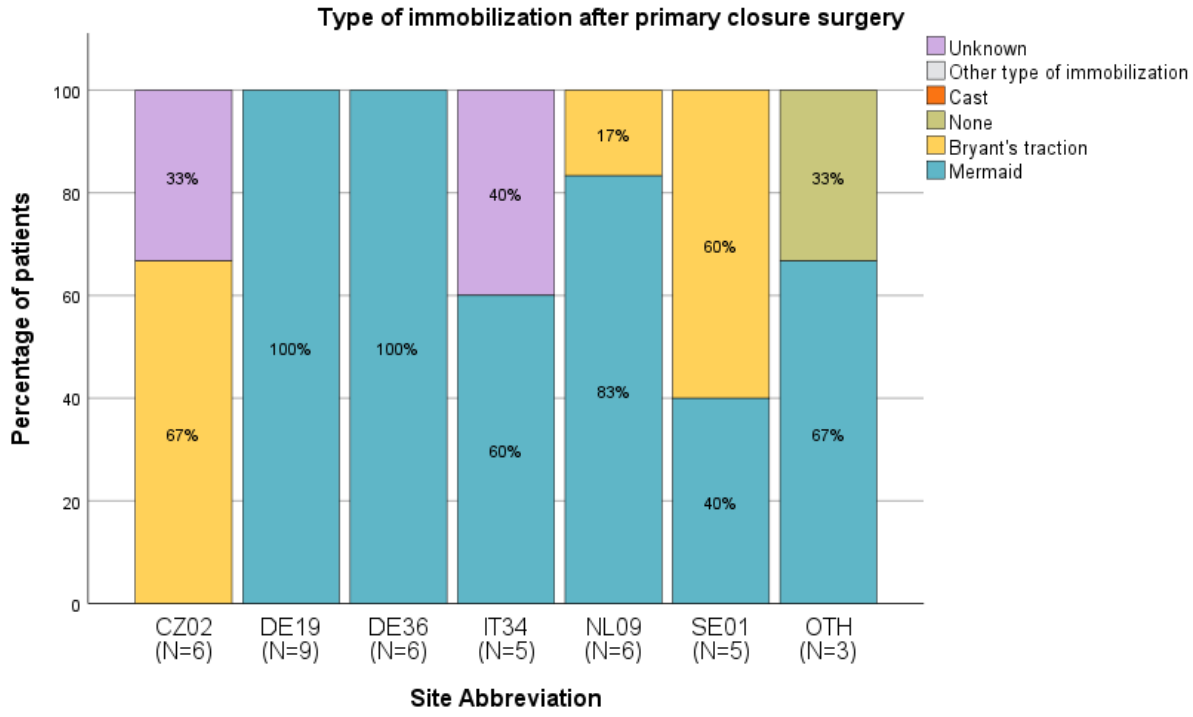
Age at primary closure

The primary closure surgery took place within 16 weeks from birth for most patients, with a peak during the first week of life.



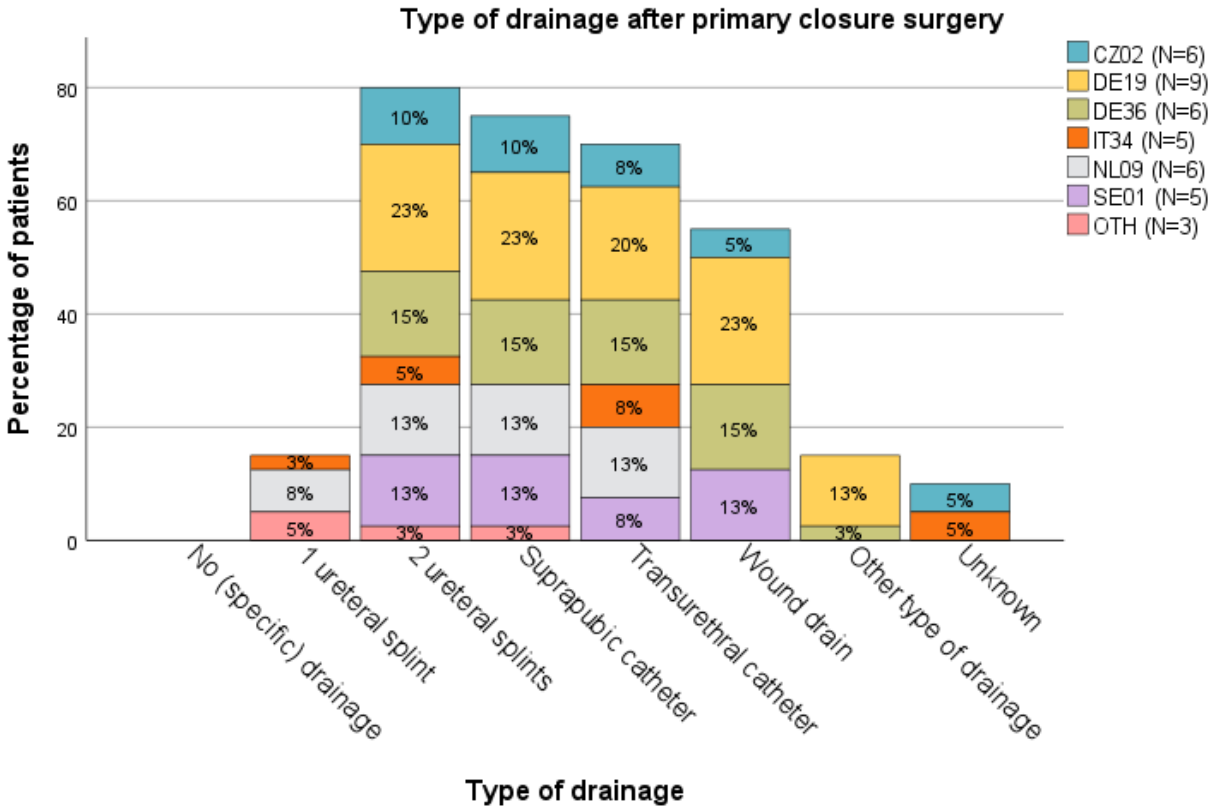
Immobilization after primary closure surgery

After surgery, the most used type of immobilization was mermaid. Bryant's traction was also used frequently across different HCPs.



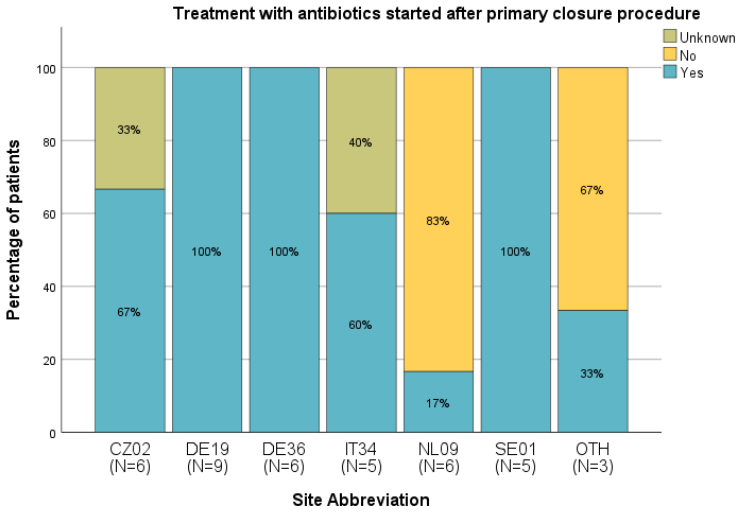
Type of drainage after primary closure surgery

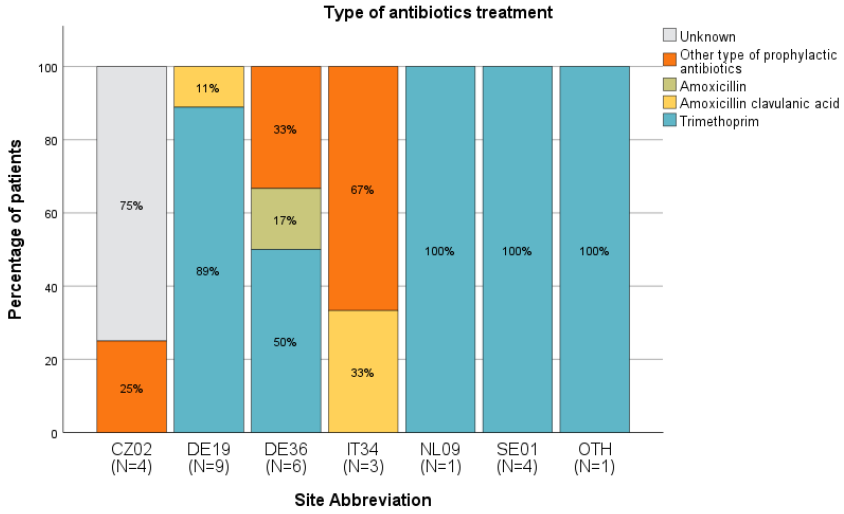
After surgery, most patients received multiple drains, with the most used type of drain being 2 urethral splints. The suprapubic and transurethral catheters were also used in more than 60% of patients. Numbers add up to more than 100% because most patients had more than one type of drainage.



Antibiotics

After primary closure surgery, most patients were started on antibiotics. Trimethoprim was used most often.

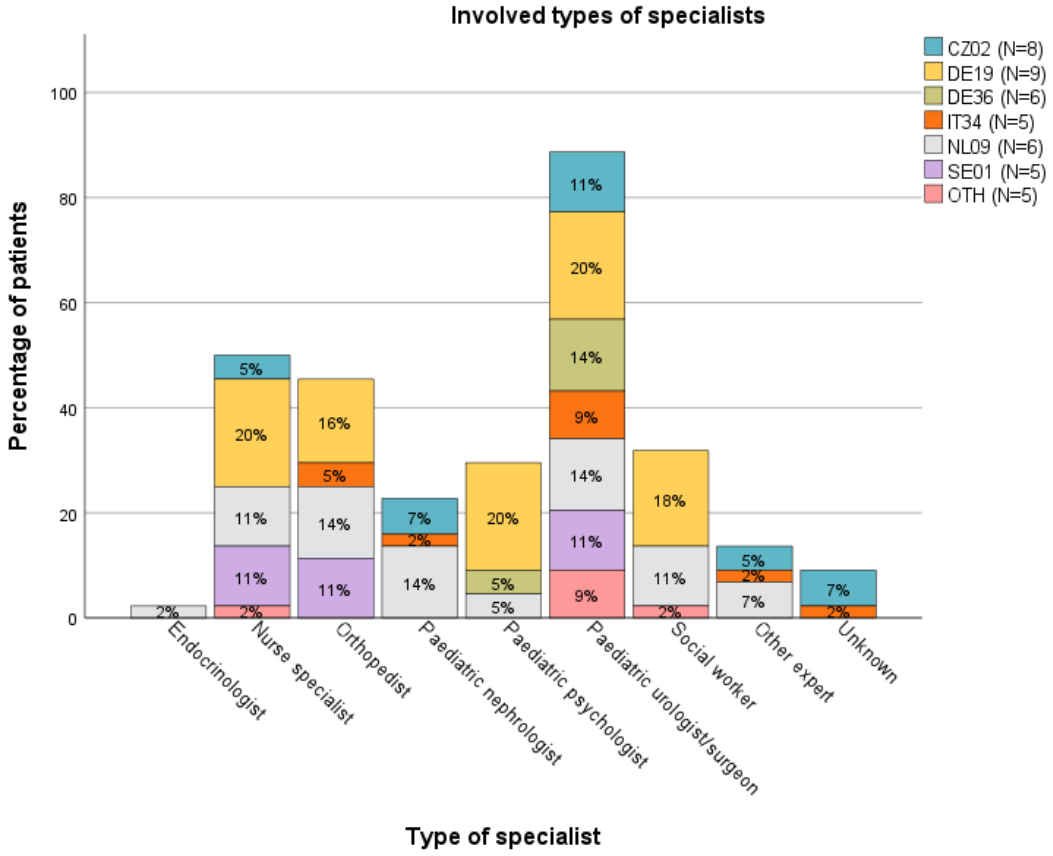




Involved specialists

Involved types of specialists

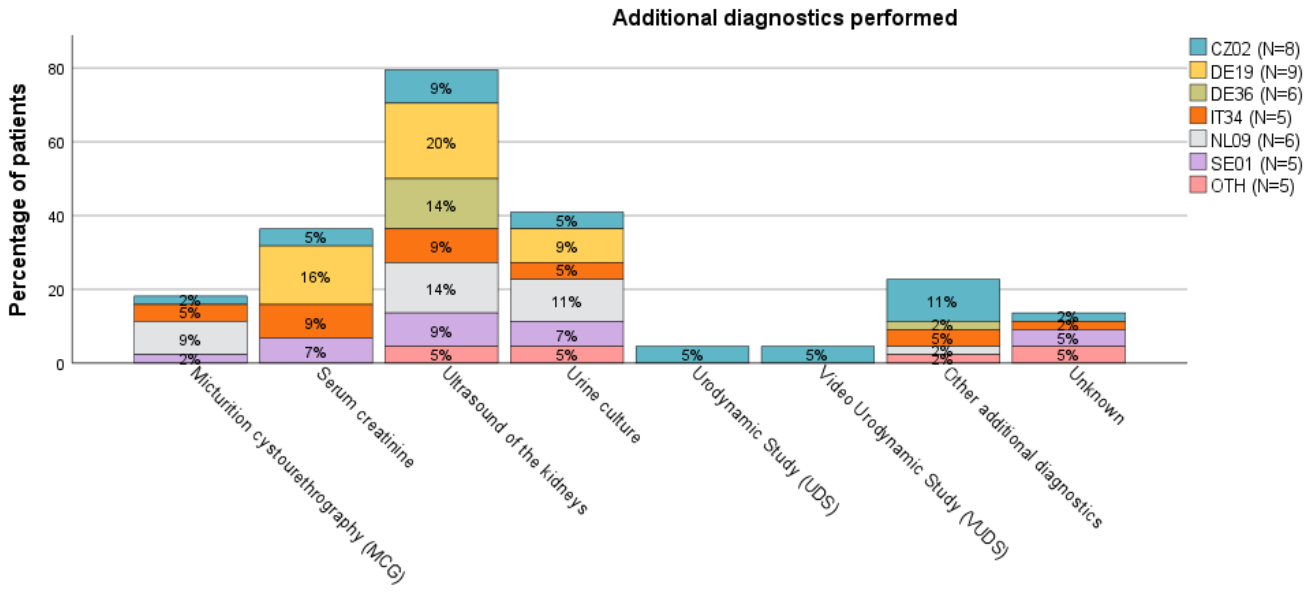
In most cases, multiple specialists were involved. A paediatric urologist/surgeon was involved in the treatment of almost all patients. Other frequently involved specialists are the nurse specialist and orthopedist. Numbers add up to more than 100% because some patients had more than one type of involved specialist.



Additional diagnostics

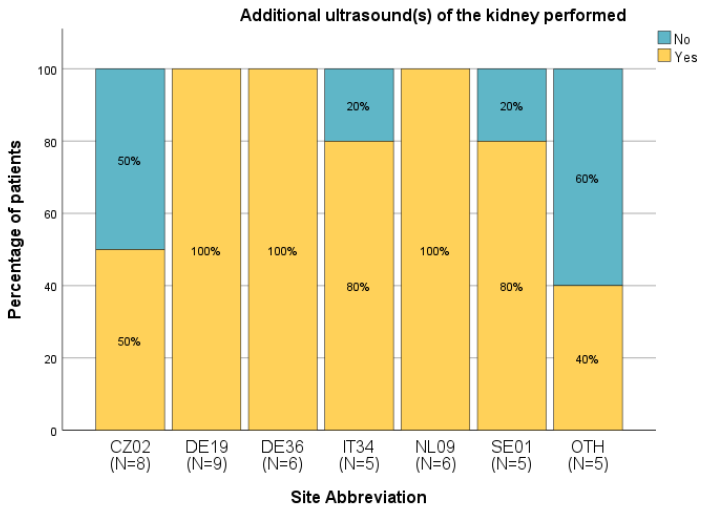
Additional diagnostics performed

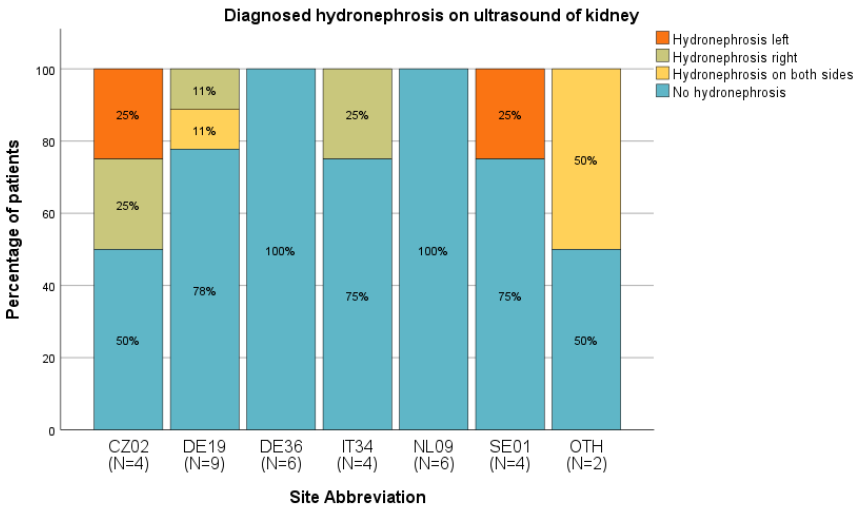
In all patients, additional diagnostics were performed during the first two years of treatment. An ultrasound of the kidneys was performed most, with 80% of patients receiving at least one. Next, urine cultures and serum creatinine were performed in approximately 40% of patients. Numbers add up to more than 100% because some patients had more than one type of additional diagnostics.



Additional ultrasounds and hydronephrosis

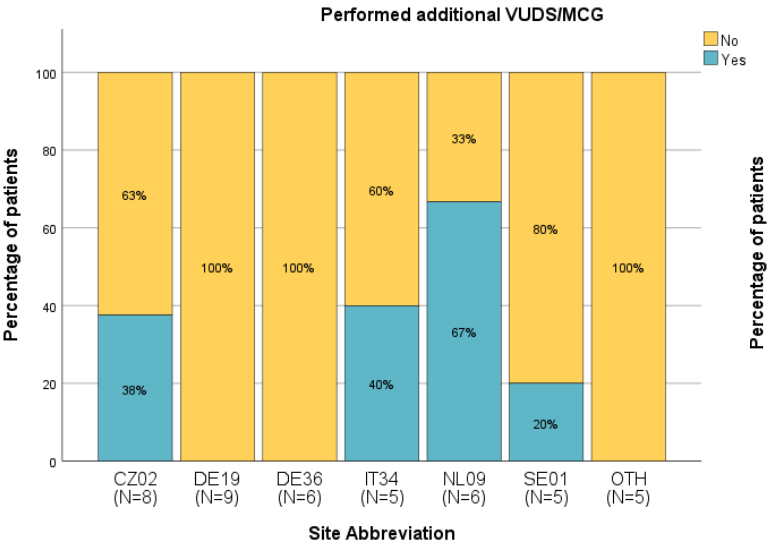
As can be seen in the previous figure, ~80% of patients received an ultrasound of the kidney. One of the outcomes of an ultrasound is hydronephrosis. In most cases, hydronephrosis was not diagnosed.

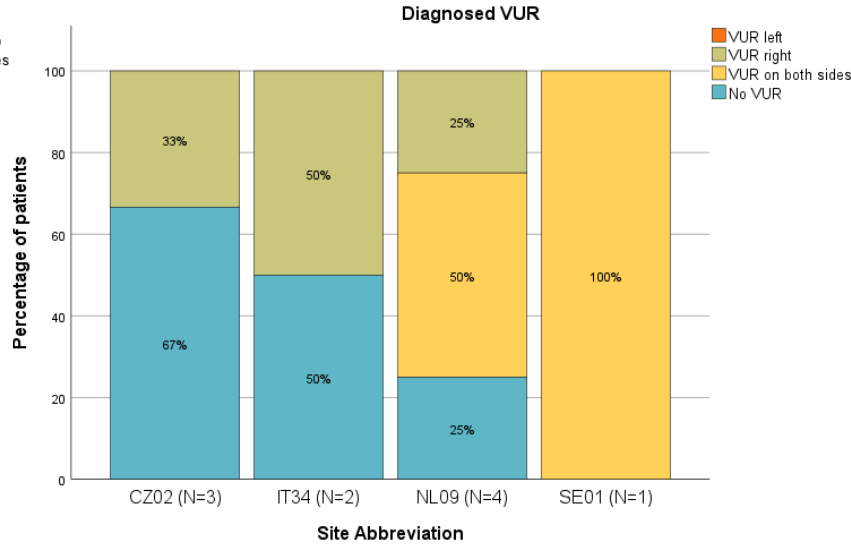




Additional VUDS/MCG and vesico-urethral reflux (VUR)

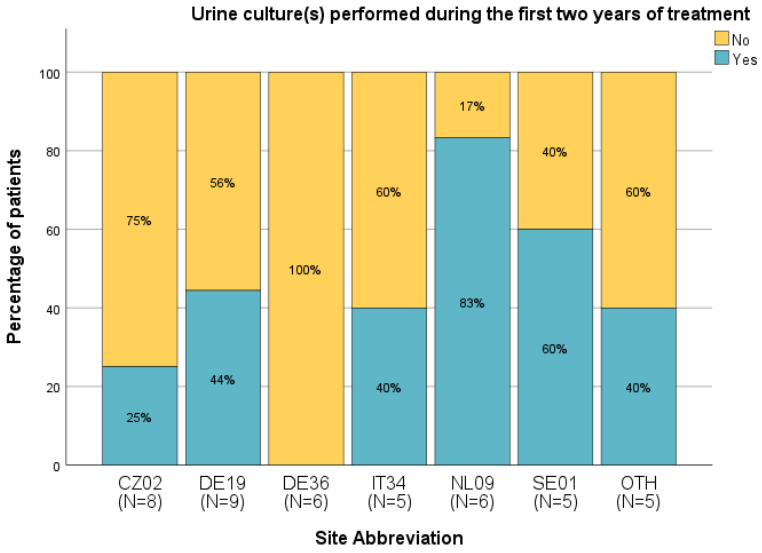
Only a small part of the patients received an additional MCG or VUDS. With both procedures, VUR can be diagnosed. In the cases where a MCG/VUDS was performed, a majority received a VUR diagnosis, with an equal amount of VUR on both sides and VUR on the right side only.

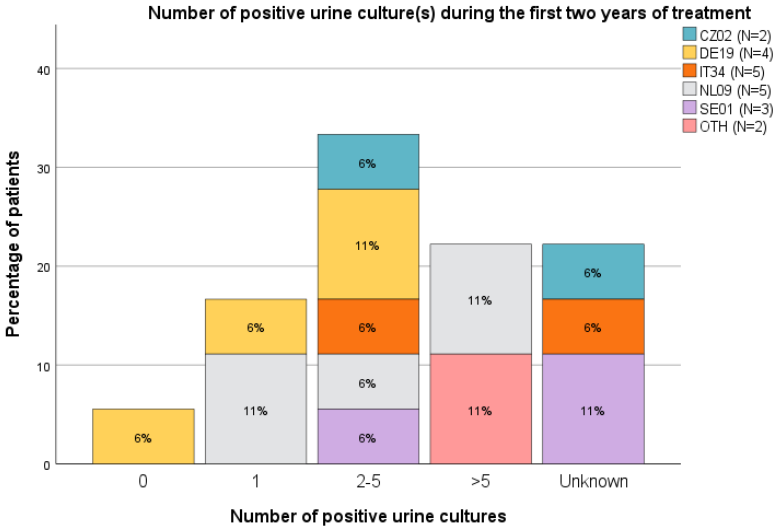




Urine cultures

Zooming in on the urine cultures, about 40% of patients received one or multiple urine cultures during the first two years of treatment. Most patients had between 2 and 5 positive urine cultures. Only a few patients had only negative urine cultures.





European Reference Networks

European Reference Network
for rare or low prevalence complex diseases

Network Urogenital Diseases (ERN eUROGEN)

http://eurogen-ern.eu/

Funded by the European Union

ERN eUROGEN is one of the 24 European Reference Networks (ERNs) approved by the ERN Board of Member States. The ERNs are co-funded by the European Commission. For more information about the ERNs and the EU health strategy, please visit http://ec.europa.eu/health/ern