

ERN eUROGEN registry

Report on Expertise Area 1.2

INTRODUCTION

This report entails an analysis of the data on Expertise Area 1.2 Classic Bladder Exstrophy 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 data of HCPs that entered more than 5 patients for 1.2 Classic Bladder Exstrophy 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: DE13 (N=2), DE22 (N=2), ES17 (N=4), IT58 (N=1). As patient numbers are not similar, 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, and we interpreted it as 'No ultrasound performed during the first two years of treatment'.

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 analyses. 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 1.2 Classic Bladder Exstrophy. Corresponding figures were made of the variables, and they are displayed on the next pages.

	Total N=78	Czechia CZ02 (N=11)	Germany		Italy IT34 (N=12)	The Netherlands NL09 (N=10)	Sweden SE01 (N=9)	Others OTH (N=9)
			DE19 (N=17)	DE36 (N=10)				
General characteristics								
Birth sex								
Male; N (%)	46 (59.0%)	10 (90.9%)	13 (76.5%)	7 (70.0%)	5 (41.7%)	3 (30.0%)	4 (44.4%)	4 (44.4%)
Female; N (%)	32 (41.0%)	1 (9.1%)	4 (23.5%)	3 (30.0%)	7 (58.3%)	7 (70.0%)	5 (55.6%)	5 (55.6%)
Primary diagnosis and treatment								
Initial wound coverage								
No coverage; N (%)	2 (2.6%)	-	-	-	1 (8.3%)	-	-	1 (11.1%)
Diaper; N (%)	15 (19.2%)	-	-	7 (70%)	1 (8.3%)	5 (50.0%)	2 (22.2%)	-
Wet gauze; N (%)	44 (56.4%)	4 (36.4%)	15 (88.2%)	7 (70%)	8 (66.7%)	-	6 (66.7%)	4 (44.4%)
Other; N (%)	15 (19.2%)	-	1 (5.9%)	9 (100%)	-	1 (10.0%)	3 (33.3%)	1 (11.1%)
Unknown; N (%)	18 (23.1%)	7 (63.6%)	1 (5.9%)	-	3 (25.0%)	4 (40.0%)	-	3 (33.3%)
Initial diagnostics								
Creatinine measurement child; N (%)	34 (43.6%)	2 (18.2%)	9 (52.9%)	5 (50.0%)	9 (75.0%)	3 (30.0%)	3 (33.3%)	3 (33.3%)
Culture bladder plate; N (%)	4 (5.1%)	-	-	-	-	4 (40.0%)	-	-
Ultrasound of kidneys; N (%)	63 (80.8%)	6 (54.5%)	17 (100%)	10 (100%)	9 (75.0%)	6 (60.0%)	9 (100%)	6 (66.7%)
X-ray of pelvis; N (%)	26 (33.3%)	-	1 (5.9%)	-	4 (33.3%)	7 (70.0%)	9 (100%)	5 (55.6%)
Other diagnostics/imaging; N (%)	24 (30.8%)	-	14 (82.4%)	-	5 (41.7%)	1 (10.0%)	2 (22.2%)	2 (22.2%)
Unknown; N (%)	13 (16.7%)	5 (45.5%)	-	1 (10.0%)	3 (25.0%)	2 (20.0%)	-	2 (22.2%)
Primary closure surgery								
Primary closure surgery performed; N (%)	73 (93.6%)	9 (81.8%)	17 (100%)	10 (100%)	12 (100.0%)	9 (90%)	9 (100%)	7 (66.7%)
Procedures at primary closure surgery								
Primary closure only; N (%)	38 (52.1%)	9 (100%)	10 (58.8%)	7 (70.0%)	6 (50.0%)	-	4 (44.4%)	2 (28.7%)
Primary closure with osteotomy; N (%)	21 (28.8%)	-	-	-	5 (41.7%)	8 (88.9%)	5 (55.6%)	3 (42.9%)
Bladder neck reconstruction; N (%)	8 (11.0%)	-	-	-	7 (58.3%)	-	-	1 (14.3%)
Epispadias reconstruction; N (%)	41 (56.2%)	2 (22.2%)	13 (76.5%)	9 (90%)	3 (25.0%)	7 (77.8%)	4 (44.4%)	3 (42.9%)
Genital mobilization in female; N (%)	9 (12.3%)	-	3 (17.6%)	1	2 (16.7%)	1	-	2 (28.7%)
Inguinal hernia repair; N (%)	20 (27.4%)	-	11 (64.7%)	6 (60.0%)	-	2 (22.2%)	1 (11.1%)	-
Introitus plasty; N (%)	5 (6.8%)	-	-	-	2 (16.7%)	1	-	2 (28.7%)
Soft in-tissue mobilization; N (%)	4 (5.5%)	-	-	-	4 (33.3%)	-	-	-
Symphysis approximation without osteotomy; N (%)	46 (63.0%)	8 (88.9%)	17 (100%)	10 (100%)	5 (41.7%)	-	4 (44.4%)	2 (28.7%)
Umbilicus reconstruction; N (%)	42 (57.5%)	2 (22.2%)	15 (88.2%)	10 (100%)	-	7 (77.8%)	6 (66.7%)	2 (28.7%)
Ureteral reimplant; N (%)	7 (9.6%)	-	-	-	3 (25.0%)	2 (22.2%)	-	2 (28.7%)
Other primary surgical procedure; N (%)	8 (11.0%)	1 (11.1%)	2 (11.8%)	4 (40.0%)	1 (8.3%)	-	-	-
Unknown; N (%)	2 (2.7%)	-	-	-	1 (8.3%)	1 (11.1%)	-	-

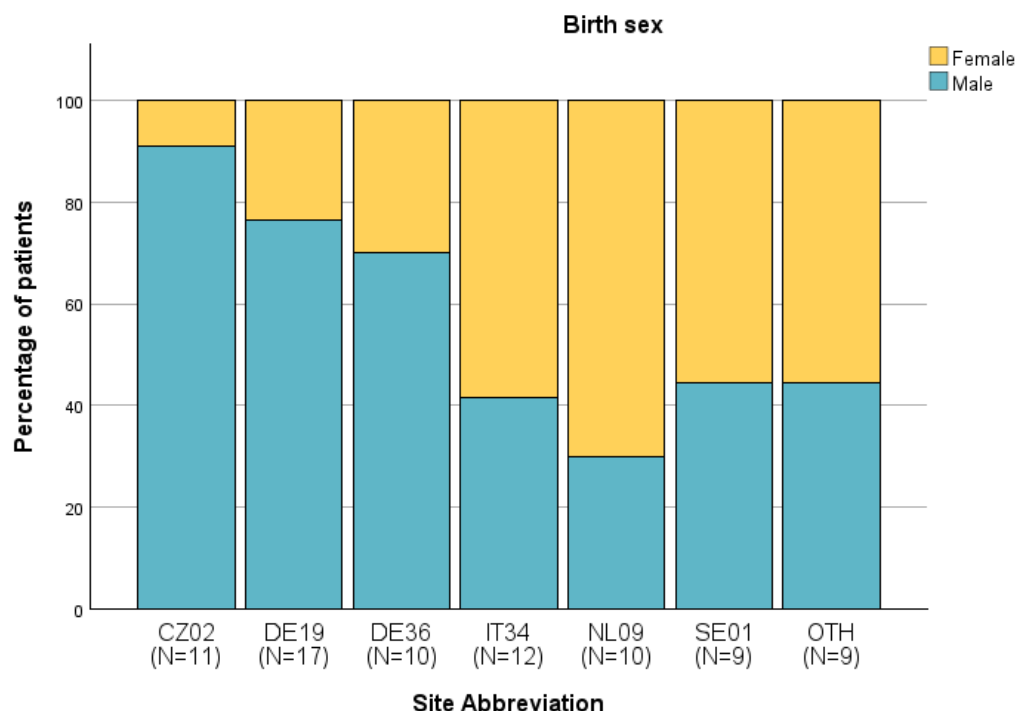
	Total N=78	Czechia CZ02 (N=11)	Germany		Italy IT34 (N=12)	The Netherlands NL09 (N=10)	Sweden SE01 (N=9)	Others OTH (N=9)
			DE19 (N=17)	DE36 (N=10)				
Age at primary closure surgery; Median (range, days)	48 days (1;9959)	2 days (1;270)	50 days (2;69)	78 days (41;110)	5 days (1;173)	10 days (5;270)	67 days (32;92)	40 days (2;9959)
Type of immobilization								
None; N (%)	1 (1.4%)	-	-	-	-	-	-	1 (14.3%)
Mermaid; N (%)	48 (65.8%)	-	16 (94.1%)	10 (100%)	7 (58.3%)	6 (66.7%)	4 (44.4%)	5 (71.4%)
Cast	2 (2.7%)	-	-	-	1 (8.3%)	-	-	1 (14.3%)
Bryant's traction; N (%)	14 (19.2%)	7 (77.8%)	-	-	-	2 (22.2%)	5 (55.6%)	-
Unknown; N (%)	8 (11.0%)	2 (22.2%)	1 (5.9%)	-	4 (33.3%)	-	-	-
						1 (11.1%)		
Post-surgical treatment								
Type of drainage								
Suprapubic catheter; N (%)	54 (74.0%)	6 (66.7%)	16 (94.1%)	10 (100%)	1 (8.3%)	7 (77.8%)	9 (100%)	5 (71.4%)
1 ureteral splint; N (%)	11 (15.1%)	1 (11.1%)	1 (5.9%)	1 (10.0%)	1 (8.3%)	3 (33.3%)	-	4 (57.1%)
2 ureteral splints; N (%)	60 (82.2%)	6 (66.7%)	16 (94.1%)	10 (100%)	7 (58.3%)	7 (77.8%)	9 (100%)	5 (71.4%)
Transurethral catheter; N (%)	52 (71.2%)	4 (44.4%)	16 (94.1%)	10 (100%)	8 (66.7%)	6 (66.7%)	4 (44.4%)	4 (57.1%)
Deep wound drain; N (%)	37 (50.7%)	2 (22.2%)	16 (94.1%)	10 (100%)	-	-	9 (100%)	-
Subcutaneous wound drain; N(%)	23 (31.5%)	-	16 (94.1%)	7 (70.0%)	-	-	-	-
Other type of drainage; N (%)	2 (2.7%)	-	2 (11.8%)	-	-	-	-	-
Unknown; N (%)	8 (11.0%)	3 (33.3%)	-	-	4 (33.3%)	1 (11.1%)	-	-
Post-surgical antibiotics treatment; N (%)	57 (78.1%)	6 (66.7%)	17 (100%)	10 (100%)	8 (66.7%)	2 (22.2%)	9 (100%)	5 (71.4%)
Type of antibiotic treatment								
Trimethoprim; N (%)	33 (57.9%)	-	16 (94.1%)	6 (60.0%)	-	1 (50.0%)	8 (88.9%)	2 (40.0%)
Nitrofurantoin; (N%)	2 (3.5%)	-	-	-	-	-	1 (11.1%)	1 (20.0%)
Amoxicillin clavulanic acid; N (%)	1 (1.8%)	-	-	1 (10.0%)	-	-	-	-
Amoxicillin; N (%)	5 (8.8%)	1 (16.7%)	1 (5.9%)	-	3 (37.5%)	-	-	-
Other type; N (%)	12 (21.1%)	2 (33.3%)	-	3 (30.0%)	5 (62.5%)	1 (50.0%)	-	1 (20.0%)
Unknown; N (%)	4 (7.0%)	3 (50.0%)	-	-	-	-	-	1 (20.0%)
			-					
Involved specialists								
Type of involved specialist								
Endocrinologist; N (%)	1 (1.3%)	-	-	-	-	1 (10.0%)	-	-
Nurse specialist; N (%)	41 (52.6%)	4 (36.4%)	17 (100%)	-	-	7 (70.0%)	9 (100%)	4 (44.4%)
Orthopedist; N (%)	40 (51.3%)	1 (9.1%)	15 (88.2%)	-	6 (50.0%)	9 (90.0%)	8 (88.9%)	1 (11.1%)
Paediatric nephrologist; N (%)	19 (24.4%)	5 (45.5%)	-	1 (10.0%)	4 (33.3%)	7 (70.0%)	1 (11.1%)	1 (11.1%)
Paediatric psychologist; N (%)	25 (32.1%)	-	17 (100%)	4 (40.0%)	-	2 (20.0%)	1 (11.1%)	1 (11.1%)
Paediatric urologist/surgeon; N (%)	71 (91.0%)	7 (63.6%)	17 (100%)	10 (100%)	11 (91.7%)	9 (90.0%)	9 (100%)	8 (88.9%)
Social worker; N (%)	24 (30.8%)	-	16 (94.1%)	-	-	5 (50.0%)	2 (22.2%)	1 (11.1%)
Other specialist; N (%)	12 (15.4%)	2 (18.2%)	3 (17.6%)	-	1 (8.3%)	4 (40.0%)	1 (11.1%)	1 (11.1%)
Unknown; N (%)	6 (7.7%)	4 (36.4%)	-	-	1 (8.3%)	1 (10.0%)	-	-

	Total N=78	Czechia CZ02 (N=11)	Germany		Italy IT34 (N=12)	The Netherlands NL09 (N=10)	Sweden SE01 (N=9)	Others OTH (N=9)
			DE19 (N=17)	DE36 (N=10)				
Additional diagnostics								
Additional diagnostics performed; N (%)	78 (100%)	11 (100%)	17 (100%)	10 (100%)	12 (100%)	10 (100%)	9 (100%)	9 (100%)
Micturition cystourethrography (MCG) ; N (%)	14 (17.9%)	3 (27.3%)	-	-	4 (33.3%)	5 (50.0%)	2 (22.2%)	-
Serum creatinine; N (%)	33 (42.3%)	3 (27.3%)	13 (76.5%)	-	7 (58.3%)	-	8 (88.9%)	2 (22.2%)
Ultrasound of kidneys; N (%)	63 (80.8%)	6 (54.5%)	17 (100%)	7 (70.0%)	10 (83.3%)	8 (80.0%)	9 (100%)	6 (66.7%)
Urine culture; N (%)	33 (42.3%)	3 (27.3%)	9 (52.9%)	1 (10.0%)	4 (33.3%)	7 (70.0%)	7 (77.8%)	2 (22.2%)
Urodynamic study (UDS) ; N (%)	4 (5.1%)	3 (27.3%)	-	-	-	-	1 (11.1%)	-
Video urodynamic study (VUDS) ; N (%)	1 (1.3%)	1 (9.1%)	-	-	-	-	-	-
Other additional diagnostics; N (%)	17 (21.8%)	6 (54.5%)	1 (5.9%)	1 (10.0%)	1 (8.3%)	4 (40.0%)	2 (22.2%)	2 (22.2%)
Unknown; N (%)	13 (16.7%)	2 (18.2%)	-	4 (40.0%)	2 (16.7%)	2 (20.0%)	1 (11.1%)	2 (22.2%)
Diagnosed hydronephrosis; N (%)	9 (14.3%)	2 (33.3%)	3 (17.6%)	-	2 (20.0%)	-	1 (11.1%)	1 (16.7%)
Diagnosed VUR; N (%)	11 (70.0%)	3 (75.0%)	-	-	3 (75.0%)	4 (80.0%)	2 (100%)	-
Urine cultures performed; N (%)	33 (42.3%)	3 (27.3%)	9 (52.9%)	1 (10.0%)	4 (33.3%)	7 (70.0%)	7 (77.8%)	2 (22.2%)
0 positive cultures; N (%)	2 (6.1%)	-	2 (22.2%)	-	-	-	-	-
1 positive culture; N (%)	7 (21.2%)	-	3 (33.3%)	-	1 (25.0%)	2 (28.6%)	1 (14.3%)	-
2-5 positive cultures; N (%)	10 (30.3%)	2 (66.7%)	4 (44.4%)	-	2 (50.0%)	1 (14.3%)	1 (14.3%)	-
> 5 positive cultures; N (%)	6 (18.2%)	-	-	-	-	3 (42.9%)	1 (14.3%)	2 (100%)
Unknown; N (%)	8 (24.2%)	1 (33.3%)	-	1 (100%)	1 (25.0%)	1 (14.3%)	4 (57.1%)	-

General characteristics

Birth sex

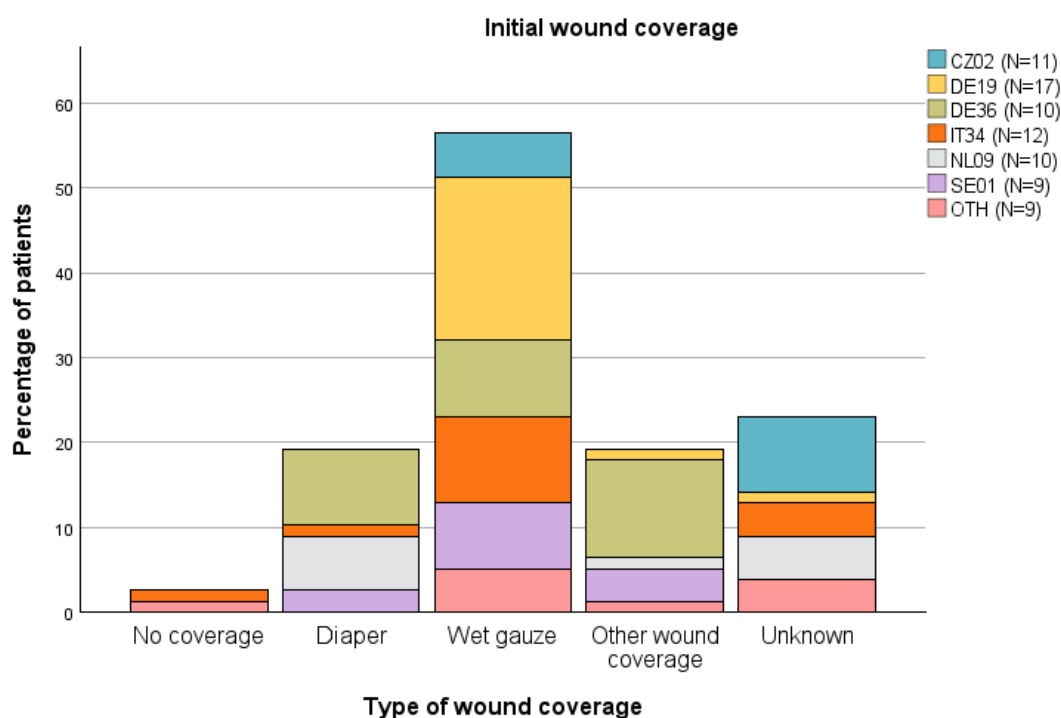
Among the registered patients, more males than females were included, although some HCPs registered more females than males.



Primary diagnosis and treatment

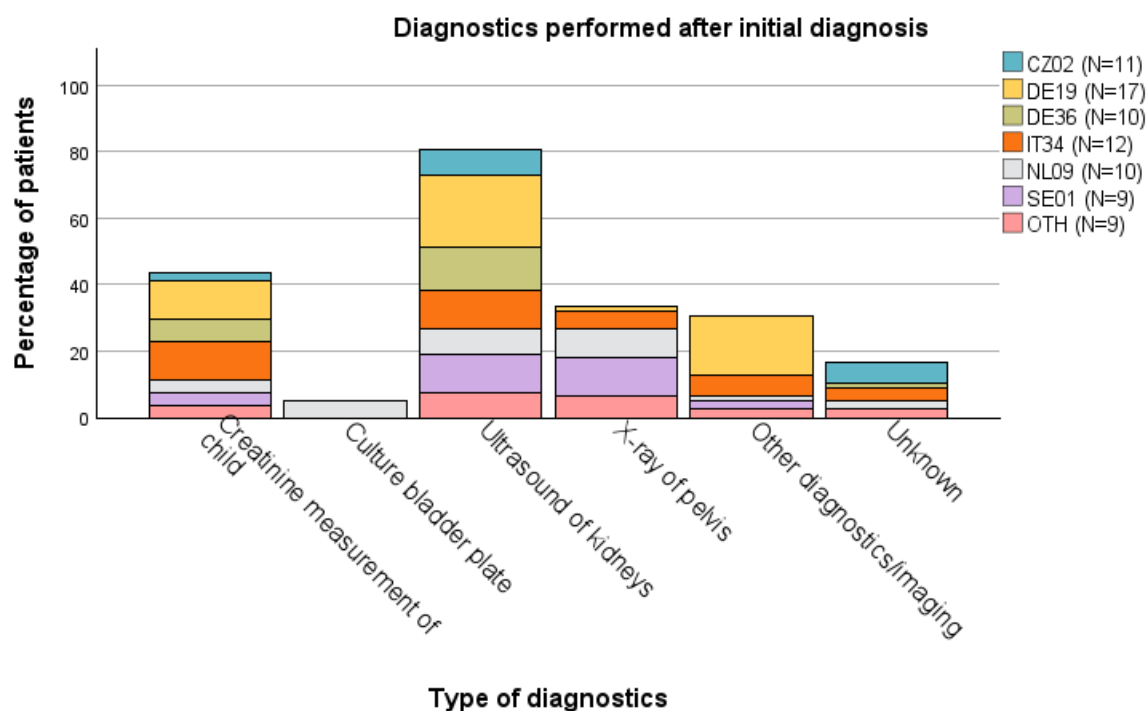
Initial wound coverage

Over half of the patients had wet gauze as initial wound coverage. Diapers and other wound coverage products were used in approximately 20% of patients. Some patients did not have any initial wound coverage.



Diagnostics performed after initial diagnosis

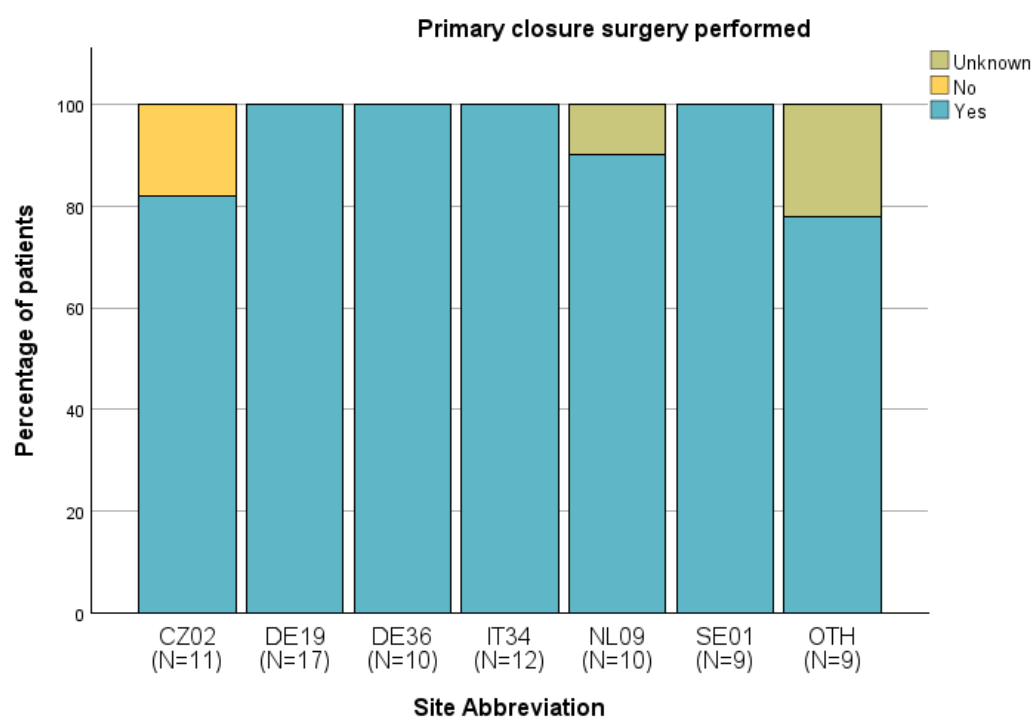
Almost all patients received an ultrasound of the kidneys during the initial diagnosis process, whereas a culture bladder plate was only performed in a small number of patients.



Primary closure surgery

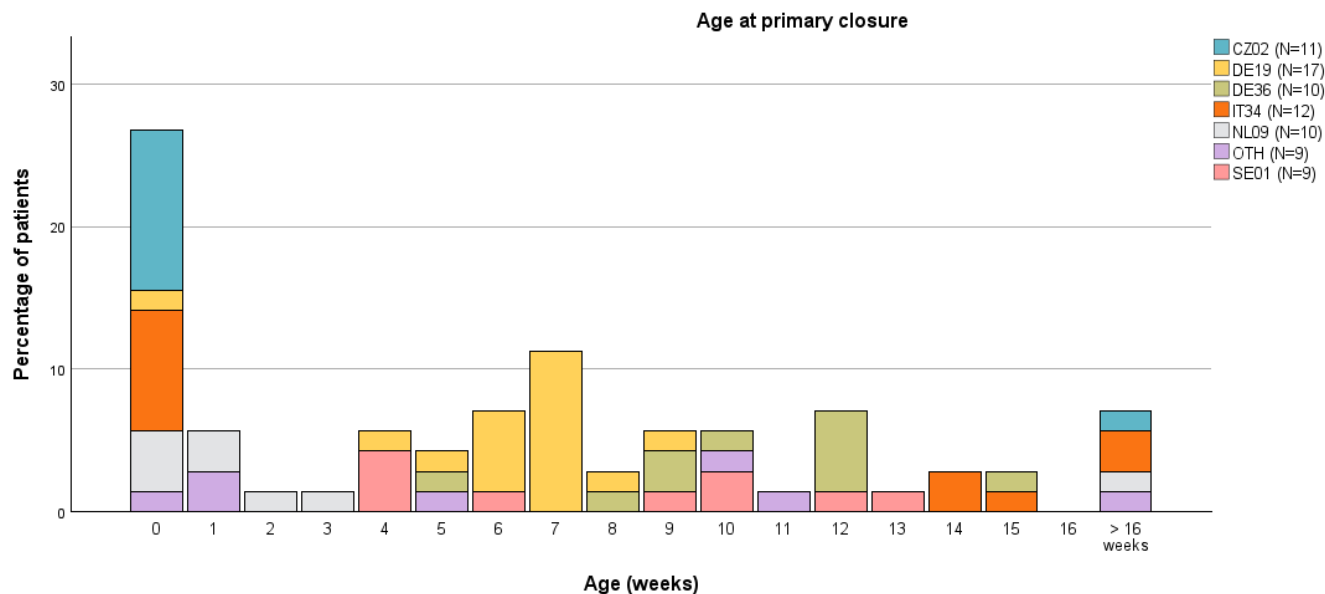
Primary closure surgery performed and location of primary closure surgery

All but a few patients had a primary closure surgery during their first two years of treatment.



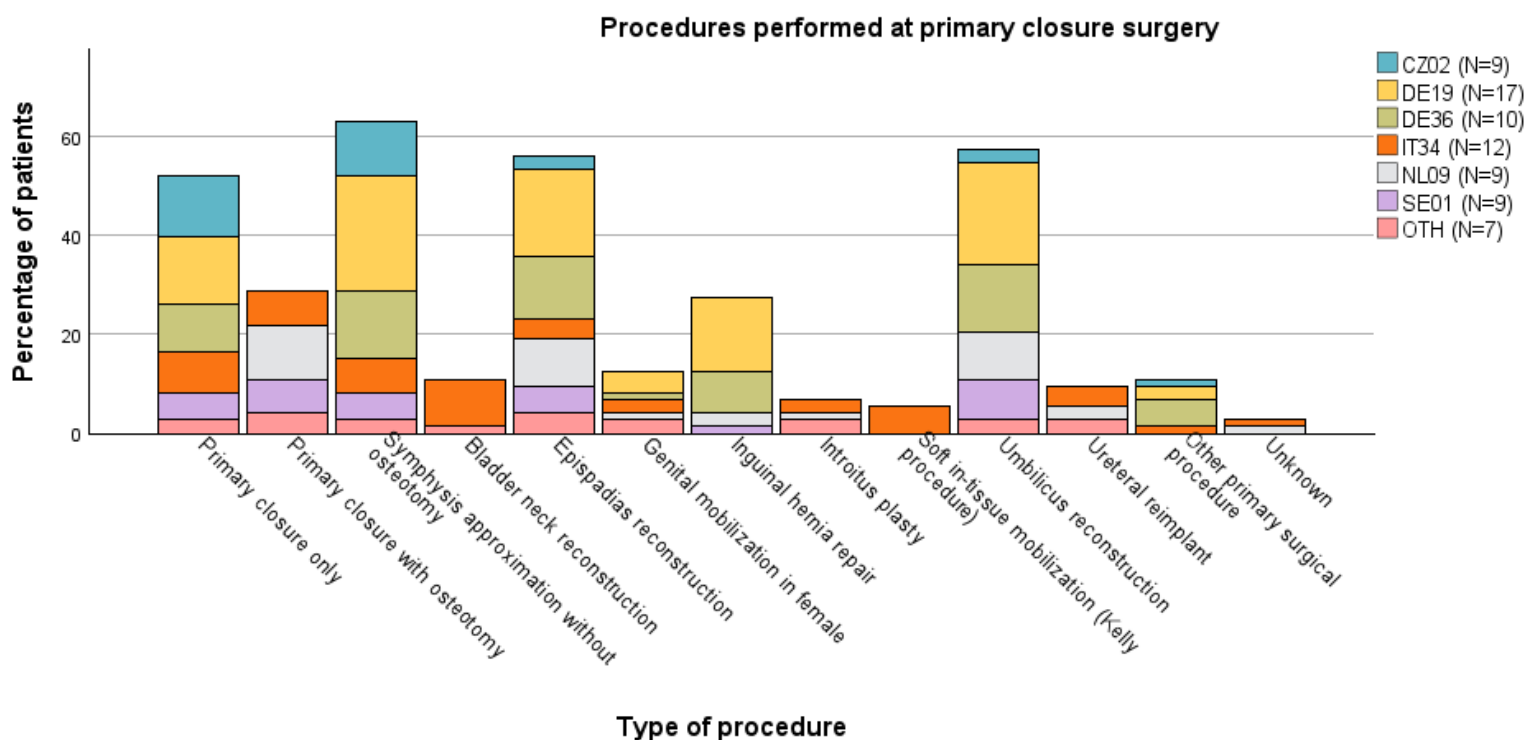
Age at primary closure

The primary closure surgery took place within 16 weeks from birth, with a peak during the first week of life. The remaining surgeries were distributed over weeks 2-16 of life.



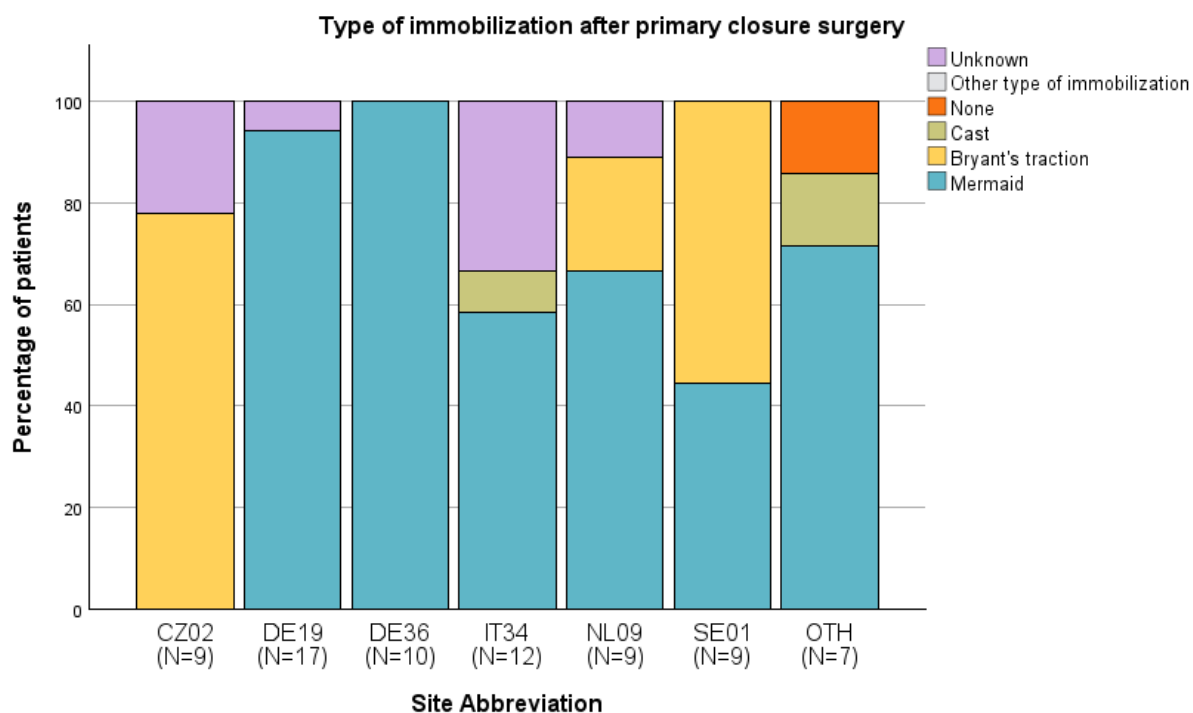
Procedures performed at primary closure surgery

At the primary closure surgery, many procedures can be performed. Most common are the symphysis approximation without osteotomy, umbilicus reconstruction and epispadias reconstruction. Introitus plasty and soft-in-tissue mobilization were only performed in a minimum of patients.



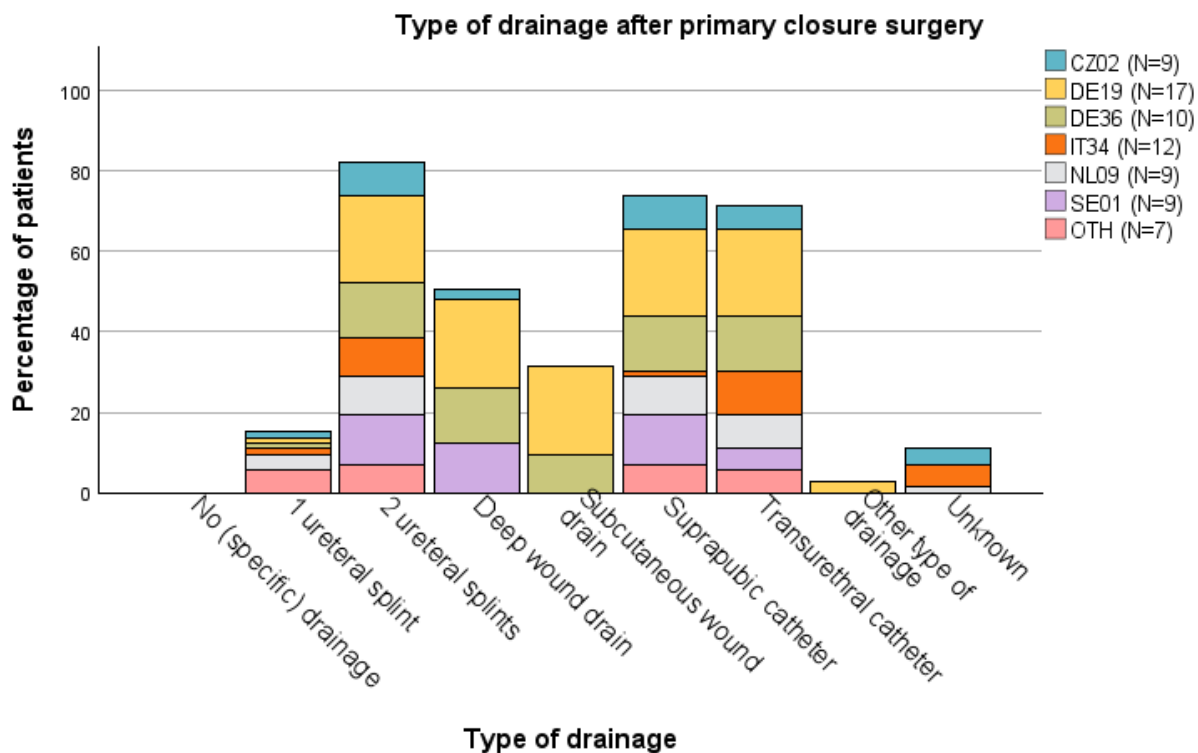
Immobilization after primary closure surgery

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



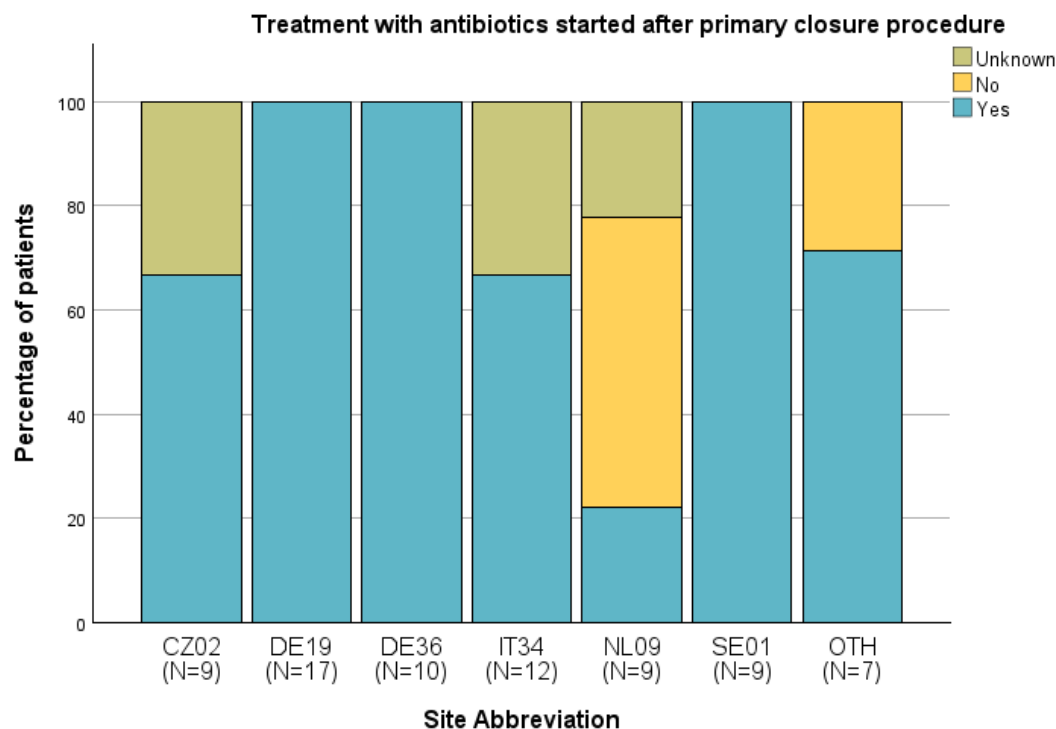
Type of drainage after primary closure surgery

After surgery, most patients received multiple drains, with the most used type of drain being 2 urethral splints. The suprapubic and transurethral catheters were used in more than 60% of patients.



Antibiotics

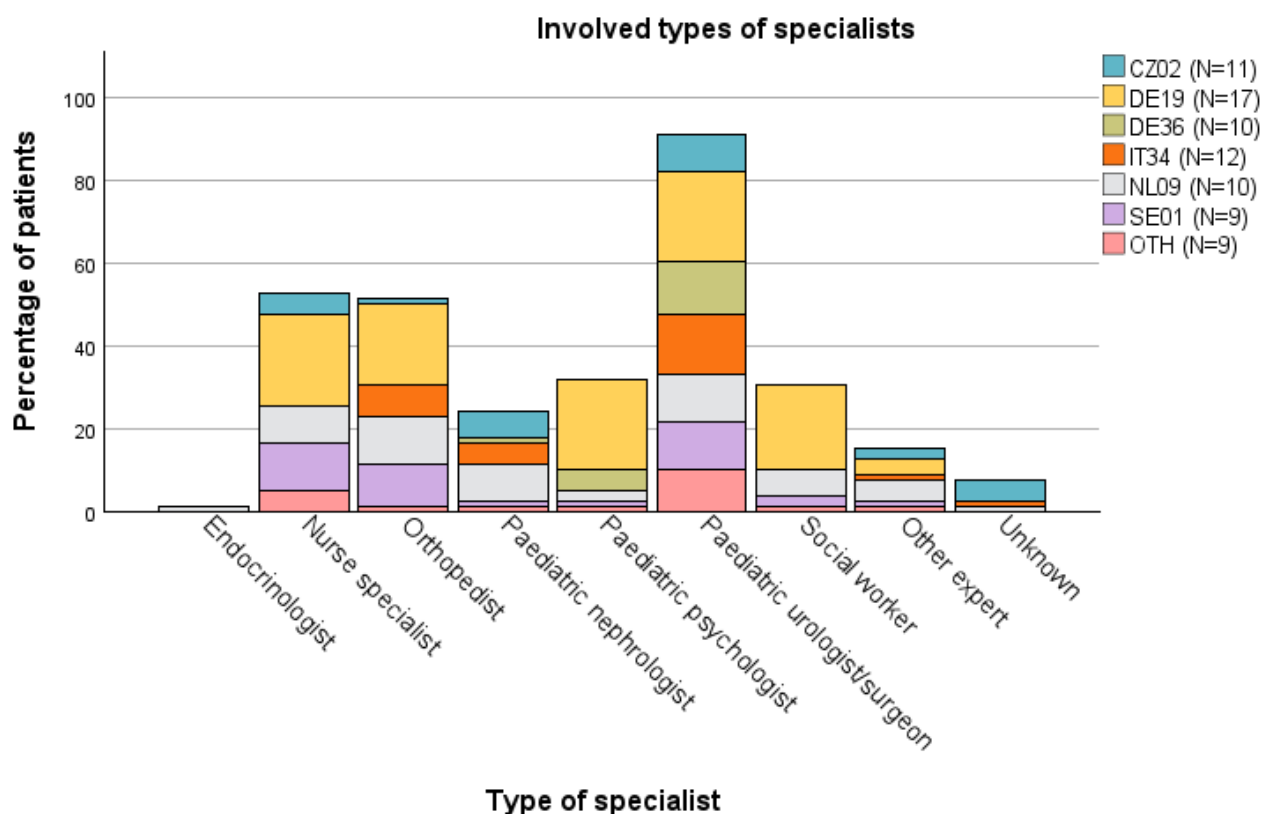
After primary closure surgery, most patients were started on antibiotics.



Involved specialists

Involved types of specialists

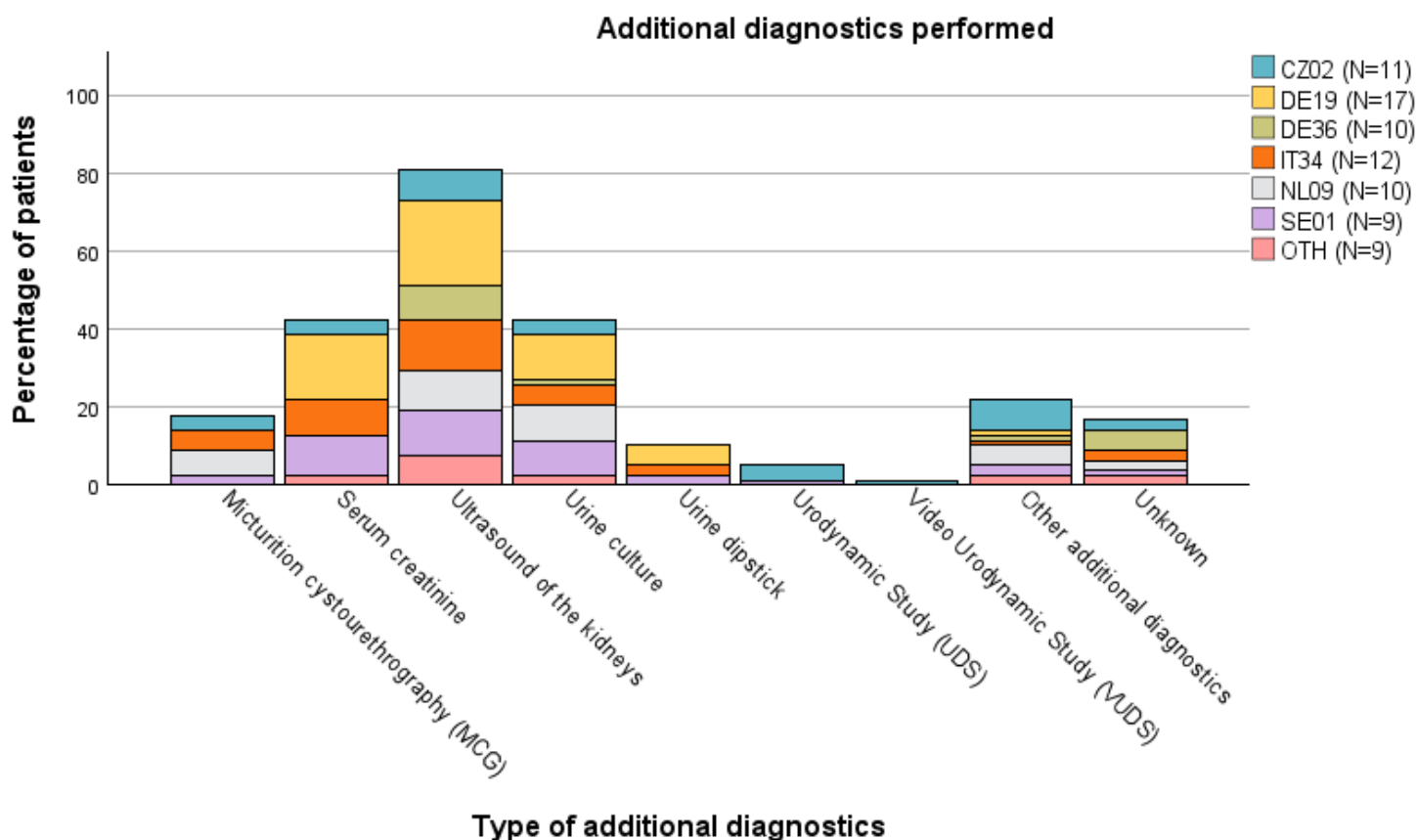
In almost all cases, a paediatric urologist/surgeon was involved the treatment. Other frequently involved specialists are the nurse specialist and orthopedist.



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.





Network
Urogenital Diseases
(ERN eUROGEN)

<http://eurogen-ern.eu/>



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