Chronic Kidney Disease Stage Progression in Patients Undergoing Repair of Persistent Cloaca

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Abbreviations and Acronyms

CIC = clean intermittent catheterization

CKD = chronic kidney disease

DTPA = ^{99m}technetium diethylenetriamine pentaacetic acid

GFR = glomerular filtration rate

NGB = neurogenic bladder

PSARVUP = posterior sagittal anorecto-vagino-urethroplasty

VUR = vesicoureteral reflux

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Purpose: Children born with persistent cloaca undergo complex pelvic reconstruction early in life. Long-term risks of bladder dysfunction and chronic kidney disease are well described. We report upper urinary tract outcomes and the risk of chronic kidney disease stage progression in this patient population.

Materials and Methods: We retrospectively studied a cohort of patients undergoing posterior sagittal anorecto-vagino-urethroplasty at a single institution from 2006 to 2013. Inclusion criteria consisted of complete urological care at our institution. Chronic kidney disease stage was calculated from cystatin C or nuclear medicine glomerular filtration rate.

Results: A total of 44 patients met inclusion criteria. Of the patients 12 had undergone vesicostomy or ureterostomy. A total of 19 patients had hydronephrosis, 19 had vesicoureteral reflux and 15 had a tethered spinal cord. Median length of the common channel was 3.5 cm. Median age at posterior sagittal anorecto-vagino-urethroplasty was 7.3 months. Median followup was 5.3 years. A total of 30 patients had neurogenic bladder, of whom 27 required clean intermittent catheterization and 3 had undergone vesicostomy. Of the patients 38 had stage I or II, 5 had stage III and 1 had stage IV chronic kidney disease. During followup no patient with initial stage I to III chronic kidney disease had stage progression. The patient with stage IV chronic kidney disease had a renal allograft placed at age 34 months before needing dialysis.

Conclusions: Early outcomes in patients with stage I to III chronic kidney disease demonstrate that renal function can be maintained despite a high rate of lower urinary tract dysfunction. Aggressive bladder management may help prevent progressive renal injury in this population.

Key Words: cloaca; glomerular filtration rate; renal insufficiency, chronic; urinary bladder, neurogenic

Children born with persistent cloaca undergo complex pelvic reconstruction early in life. The long-term risks of bladder dysfunction and progressive renal deterioration have been well described in patients with anorectal malformation and its variants.[1–8] At our institution a collaborative multidisciplinary approach among all providers has long been practiced to coordinate anorectal and genitourinary management. In addition, our protocol includes an early aggressive approach to the lower urinary tract with a low threshold to institute clean intermittent catheterization in the

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face of hostile bladder dynamics. We report upper urinary tract outcomes and the risk of chronic kidney disease progression in this patient population.

MATERIALS AND METHODS

After obtaining institutional review board approval we retrospectively studied all patients undergoing PSARVUP for persistent cloaca at a single institution from 2006 to 2013. The study period was selected to coincide with the arrival at our institution of the senior author (AP) and the creation of the Alberto Peña Colorectal Center for Children. Patients were excluded if they did not receive complete urological care postoperatively at our institution. Patient demographics, presence of concomitant renal anomalies, surgical and intraoperative details, treatment of lower urinary tract dysfunction and details of any subsequent lower urinary tract reconstruction were abstracted from the medical record.

Patients presenting with persistent cloaca were evaluated in a multidisciplinary fashion by the colorectal, urology and gynecology services. Urological testing included renal and bladder sonography, voiding cystourethrography, filling cystometrogram, leak point pressure determination and, often, cystoscopy with the patient under anesthesia. Neonatal spinal ultrasound and/or magnetic resonance imaging was performed in all patients to assess for the presence of spinal cord tethering. Laboratory evaluations were obtained, including serum creatinine and estimated glomerular filtration rate by measurement of cystatin C. Our practice is to include assessment of renal function by calculating GFR to determine CKD stage at least once yearly. A nuclear medicine GFR was obtained selectively at the discretion of the treating physician.

Aggressive bladder management was initiated in cases where there was concern regarding neurogenic bladder based on imaging and/or urodynamic evidence of severe bladder dysfunction. Treatment options included CIC and anticholinergic medication if indicated by bladder instability and/or increased detrusor leak point pressure. A proactive approach to start CIC was undertaken in patients who were being prepared for urinary undiversion at cloaca repair. In patients with a vesicostomy an attempt to perform CIC through the common channel was initially undertaken before vesicostomy closure.

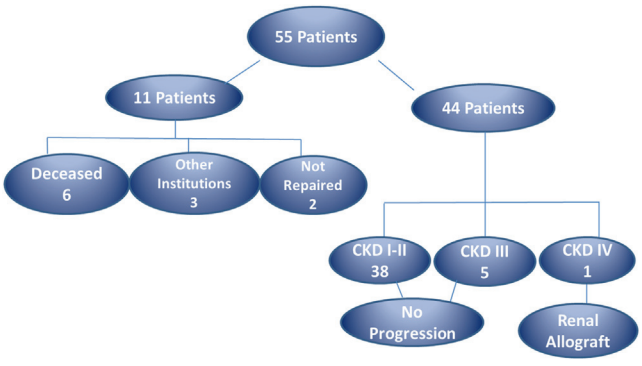
CKD stage was primarily determined using GFR estimated from serum cystatin C using the method described by Larsson et al.[9] This protocol has been validated in children and adults. GFR measured by DTPA scan was also analyzed if available. For DTPA a single component negative exponential model was used.[10] Four blood samples were obtained to allow calculation of plasma disappearance kinetics. Immediate and delayed imaging was performed. Nuclear medicine GFR was corrected for body surface area. Estimated GFR from serum creatinine (ie Schwartz formula) was not used for CKD staging purposes. However, serum creatinine values were collected in the database and analyzed to verify that they trended appropriately with the cystatin C or nuclear medicine GFR.

CKD stages were defined using the National Kidney Foundation KDOQI (Kidney Disease Outcomes Quality Initiative)™ guidelines.[11] Stage I is considered normal kidney function (GFR greater than 90 ml per minute) with findings of renal and/or bladder structural abnormalities that indicate kidney disease, stage II is mildly decreased kidney function (60 to 89), stage III is moderately reduced kidney function (30 to 59), stage IV is severely decreased kidney function (15 to 29) and stage V is very severe or end-stage kidney failure (less than 15, or patient on dialysis). Cases with increased cystatin C GFR (defined as GFR greater than 130 ml per minute for study purposes) were categorized as stage I. However, they were analyzed in detail for possible hyperfiltration injury, including assessment for proteinuria and microalbuminuria. Kaplan-Meier curves of CKD stage through time were generated and analyzed for stage progression.

RESULTS

A total of 55 patients with persistent cloaca were identified during the study period, of whom 44 met inclusion criteria. Six infants died of other major nonrenal anomalies in the first few months of life, 2 patients have not undergone repair, and 3 patients are being followed elsewhere and urological data are unavailable (see figure). A total of 27 patients (61%) were referred with a preexisting colostomy, and 12 patients had undergone urinary diversion with vesicostomy or ureterostomy before referral. On initial evaluation 19 patients had hydronephrosis and 19 had VUR (10 with grade IV or V, 7 with grade III and 2 with grade II disease), of whom 4 had bilateral VUR.

The majority of PSARVUP procedures were performed by the senior author (AP). No patient underwent redo of PSARVUP performed elsewhere. A total of 16 patients underwent 1 or more concomitant urological procedures during cloaca repair, of whom 15 underwent vesicostomy creation or revision of an existing vesicostomy. Two patients underwent nephrectomy and 1 underwent bilateral



Renal outcomes in children with persistent cloaca treated at single institution.