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Management Challenges in Pediatric Prune Belly Syndrome with Renal Failure

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Description

Prune Belly Syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare congenital disorder characterized by a classic triad: deficient or absent abdominal musculature, bilateral cryptorchidism, and urinary tract abnormalities, most notably a massively dilated bladder and ureters. Among the most serious complications of PBS is renal failure, which may be present at birth or develop progressively due to underlying uropathy and dysplasia. The management of pediatric patients with PBS and renal failure presents a range of clinical, surgical, and ethical challenges, making care complex and highly individualized. Once renal failure is identified or suspected, a multidisciplinary team approach is essential. This includes pediatric nephrologists, urologists, surgeons, and intensivists. One of the key early decisions is the management of the urinary tract. In infants with massive bladder distension and poor emptying, bladder drainage is often required. Options include Clean Intermittent Catheterization (CIC), vesicostomy, or placement of a suprapubic catheter. However, implementing CIC in a floppy, atonic bladder can be technically difficult, particularly when there is urethral or bladder neck obstruction. Vesicostomy, while often effective in decompressing the upper tracts, may not prevent the progression of renal damage in cases of severe dysplasia.

A significant surgical challenge arises in determining the timing and extent of urological reconstruction. Traditional procedures such as abdominoplasty, orchiopexy, and urinary tract reconstruction (e.g., ureteral tapering, bladder augmentation) are often delayed until the patient is medically stable. In children with End-Stage Renal Disease (ESRD), however, the timing of such interventions must be coordinated with plans for dialysis or transplantation. Reconstructive surgery carries increased risk in patients with poor nutrition, anemia, and electrolyte imbalances-all common in ESRD.

The initiation of Renal Replacement Therapy (RRT) in infants with PBS and ESRD presents another layer of complexity. Peritoneal dialysis is generally preferred in younger children due to technical feasibility and the challenge of maintaining vascular access for hemodialysis. However, PBS patients may have an increased risk of complications with peritoneal dialysis due to abdominal wall laxity and frequent urinary tract infections. Dialysate leakage, catheter dysfunction, and peritonitis are not uncommon. In some cases, temporary hemodialysis is required, which demands meticulous care in securing reliable vascular access in small infants.

Nutritional support is a cornerstone of management but also presents difficulties. Children with PBS often have feeding difficulties, failure to thrive, and protein-energy malnutrition, particularly when on dialysis. Adequate caloric intake is essential for growth, wound healing, and eventual transplant candidacy. Additionally, electrolyte abnormalities, metabolic acidosis, and anemia must be aggressively managed, often requiring

multiple medications and close monitoring.

Another key consideration is the timing of renal transplantation. While transplantation offers the best long-term survival and quality of life, PBS patients often face delays due to recurrent infections, bladder dysfunction, or poor growth. Moreover, the presence of a poorly compliant or dysfunctional bladder can compromise graft function. Urodynamic studies are frequently performed pre-transplant to evaluate bladder capacity, compliance, and voiding pressures. In cases of high-pressure or noncompliant bladders, pre-transplant bladder augmentation or urinary diversion may be required to protect the graft. These procedures carry their own risks and may further delay transplantation.

Immunological and psychosocial aspects further complicate management. Immunosuppressive therapy post-transplant increases the risk of infection, particularly in children already prone to recurrent urinary tract infections. Close coordination with infectious disease specialists is necessary to develop prophylactic strategies. Psychosocial challenges, including caregiver fatigue, chronic hospitalizations, and emotional distress, are common in families of children with PBS and renal failure. Access to psychological support and social services is essential to help families cope with the long-term demands of care. Long-term follow-up is essential for all PBS patients, particularly those who undergo transplantation. They require ongoing surveillance for graft function, growth parameters, urologic complications, and pubertal development. Male patients with PBS often have impaired fertility due to cryptorchidism and testicular dysgenesis. Hormonal evaluations and fertility counseling may be indicated during adolescence. Additionally, lifelong monitoring for cardiovascular risk factors, including hypertension and dyslipidemia, is necessary due to chronic kidney disease and immunosuppressive therapy.

Conclusion

The management of pediatric prune belly syndrome with renal failure is fraught with clinical and surgical challenges that demand a multidisciplinary, patient-centered approach. Early identification of renal dysfunction, careful urinary tract management, optimization for dialysis and transplant, and comprehensive family support are all vital components of care. As survival improves, attention must also shift toward enhancing quality of life, supporting growth and development, and preparing these children for a healthy transition into adulthood. Continued research and collaboration among centers are needed to establish standardized protocols and improve outcomes for this rare and complex condition.