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METABOLIC CONSEQUENCES AND LONG-TERM COMPLICATIONS OF ENTEROCYSTOPLASTY IN CHILDREN: A REVIEW

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ABSTRACT

Purpose: We summarize important metabolic consequences and long-term complications associated with enterocystoplasty with particular emphasis on the pediatric patient with genitourinary abnormalities.

Materials and Methods: A directed MEDLINE literature review for metabolic and long-term complications following enterocystoplasty was performed. Information gained through the published literature and from our database was reviewed and summarized to provide the reader with a thorough review of the subject.

Results: Bowel is not a perfect tissue for substitution or augmentation and its use to treat functionally and structurally compromised bladders is associated with several metabolic consequences and long-term complications. Metabolic acidosis is the most common metabolic abnormality seen. The rates and severity of these complications vary, although they may have a profound impact on patient quality of life after enterocystoplasty.

Conclusions: The metabolic consequences and long-term complications associated with enterocystoplasty are important clinical features of this intervention. Careful consideration should be given to them prior to pursuing enterocystoplasty.

KEY WORDS: bladder, urinary diversion, metabolism, gastrointestinal tract, complications

Enterocystoplasty is commonly used in children born with developmental abnormalities involving the genitourinary system and it has become a standard part of the armamentarium for the management of neurogenic and myogenic bladder. Although the principles applied are fairly standard, various techniques and bowel segments can be used depending on the clinical situation. The incorporation of bowel segments in the genitourinary tract is associated with several metabolic and mechanical consequences. The gastrointestinal tract is a relatively poor substitute for urothelium and its semipermeability permits nonphysiological fluid and electrolyte absorption, leading to metabolic abnormalities. Further complicating matters, the loss of various portions of the gastrointestinal tract can interfere with normal gastrointestinal function, resulting in complications such as chronic diarrhea, which carry additional attendant metabolic consequences. We summarize and categorize the most common metabolic abnormalities and long-term consequences encountered with enterocystoplasty.

SIGMOID

Ureterosigmoidostomy was an early and initially popular form of continent urinary diversion, particularly for the management of bladder exstrophy. High rates of hyperchloremic, hypokalemic metabolic acidosis, ascending urinary tract infection and pyelonephritis, and an increased rate of colon cancer have effectively decreased the application of ureterosigmoidostomy.[1] Many of these patients present with symptoms of metabolic acidosis, and recurrent episodes of urosepsis and pyelonephritis. At our institution all adult patients with a history of exstrophy initially managed by ureterosigmoidostomy are now counseled toward ureterosigmoidostomy reversal, creation of an alternative urinary diversion separating the urinary and fecal streams and removal of the sigmoid cuff surrounding the site of the ureteral reimplantation because of the high risk of cancer development.

ILEUM, CECUM AND COLON

The ileum and segments of the large bowel are the most commonly used components of the gastrointestinal tract for enterocystoplasty and continent urinary diversion. Advantages obtained when using ileum are the abundant supply of small bowel, a predictable and abundant mesenteric blood supply, compliance relative to that of other bowel segments, moderate mucous production compared to colon and less severe associated metabolic complications compared to stomach or colon. Ileum is not a suitable bowel segment to use in certain situations. Contraindications are a history of short gut syndrome, inflammatory bowel disease, pelvic and abdominal irradiation, and significant renal insufficiency. Although they are commonly used for bladder augmentation and urinary diversion, ileal segments are associated with several limitations. They include the occasional development of hyperchloremic, hypokalemic metabolic acidosis, vitamin B_{12} deficiency, diarrhea and steatorrhea, difficulty creating submucosal tunnels for ureteral implantation and urinary tract infection. Several of these challenging issues are addressed subsequently in greater detail.

Colon enterocystoplasty is complicated by several anatomical and functional characteristics unique to the large bowel. Shorter mesenteries can make mobilization and reconfiguration of the large bowel more difficult. Furthermore, colon is associated with a greater degree of mucous formation.

In intestinal urinary diversion the net absorption of NH_4 is the major contributor to increased acid load and metabolic acidosis.[2] To a lesser extent bicarbonate losses, the use of bicarbonate to titrate endogenous and ingested acids, and the inability of the kidneys to secrete acid contribute to acidosis.

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In patients with adequate renal function the kidneys are able to process the increased acid load and seldom do clinical consequences arise. Still, approximately 50% of patients with ileal bladder replacement have metabolic acidosis[3] and more than 50% with a colonic reservoir have some degree of measurable hyperchloremic metabolic acidosis. Ureterosigmoidostomies have even higher rates of metabolic acidosis, reaching up to 80%.[4]

There is some evidence that the absorptive properties of the intestinal segment used for enterocystoplasty may decrease with time,[5] although this is not entirely clear.[6] Certainly histological changes in the intestinal mucosa occur, including mucosal atrophy and decreased villous height,[7] and they have been implicated as a potential mechanism for decreased intestinal absorptive capacity.[8] In most patients who have undergone enterocystoplasty acidosis is typically not severe. The clinical significance of minor acidosis has not been well elucidated, although a majority of patients have measurable electrolyte abnormalities, such as increased Cl^- and NH_4^+ levels.[9] In older series hyperchloremic acidosis was reported in up to 68% of patients with an ileal conduit.[10,11] Metabolic acidosis has been reported as a major problem in 18% of patients in at least 1 older series of patients with intestinal cytoplasties.[12] Symptoms were easy fatiguability, anorexia, weight loss, polydipsia and lethargy.

Primary treatment should be alkalinization with oral sodium bicarbonate. However, other oral alkalinizing agents can be used, such as sodium citrate and citric acid solutions. These therapies are not always well tolerated because sodium bicarbonate produces considerable gas and the taste of sodium citrate can be unpalatable.[13] The use of chlorpromazine[14] or nicotinic acid[15] may limit the degree of acidosis and decrease the need for alkalinizing agents, although it does not correct the acidosis alone. The 2 agents act through inhibiting cyclic adenosine monophosphate and impeding chloride transport. The side effects can be severe, including tardive dyskinesia. The medications should not be used in patients with peptic ulcer disease or significant hepatic insufficiency.

Low serum potassium, as seen in patients with continent urinary diversion, typically results from renal potassium wasting and chronic metabolic acidosis, which causes intracellular potassium depletion. Ileal segments have a greater capacity for potassium reabsorption compared to colon segments,[16] which may help attenuate hypokalemia in patients with ileal augments and diversion. Continent colon urinary diversions and ureterosigmoidoscopies carry an increased risk of hypokalemia compared to ileal neobladders.[17] Severe depletion may result in flaccid paralysis. Treatment for hypokalemia consists of replacing potassium in addition to correcting acidosis with bicarbonate. Care must be taken during the correction of acidosis to monitor potassium levels closely since the correction of acidosis results in an intracellular potassium shift and can lead to profound hypokalemia.

Although hypocalcemia and hypomagnesemia can occur with urinary diversion, they are uncommon. Chronic metabolic acidosis results in the loss of calcium stores from bone as phosphates and sulfates are used to buffer the acids. Clinically significant hypocalcemia can result in symptoms, including tetany, tremors, irritability and in extremely severe cases death. Treatment is based on calcium repletion. Hypomagnesemia results from magnesium malabsorption, renal loss and decreased renal tubular absorption secondary to acidosis.[18] Symptoms are similar to those that occur with low calcium levels and treatment consists of exogenous replacement.

GASTROCYSTOPLASTY

The use of stomach for urinary diversion and augmentation cystoplasty is not common. However, in certain specific instances gastric segments can be a valuable adjunct to more commonly used techniques. The metabolic benefits of gastric cystoplasty are intestinal sparing and the prevention of short bowel syndrome, a decreased occurrence of hyperchloremic metabolic acidosis and decreased mucous production, translating into lower rates of urine infection and stone formation. Patients with renal insufficiency and chronic acidosis may benefit from the inclusion of a gastric segment due to the ability of the stomach to secrete acid.[19] The use of stomach in patients requiring continent urinary diversion or bladder augmentation is not without consequences. Fluid, potassium and chloride losses may develop, resulting in severe dehydration and hypochloremic, hypokalemic metabolic alkalosis. These metabolic abnormalities may precipitate clinical presentations, ranging from lethargy and mental status changes to intractable seizures. In severe cases respiratory compromise related to the compensatory respiratory acidosis that develops in response to metabolic alkalosis can be seen.[20]

Patients with urinary reconstruction including gastric segments must be instructed to maintain adequate hydration and salt intake to prevent clinically significant dehydration from developing. Acute gastrointestinal illness can precipitate severe metabolic alkalosis requiring hospitalization, particularly in patients with baseline renal insufficiency. When patients present with symptoms, electrolyte and fluid repletion are the primary treatment objectives, and normal saline infusion and potassium repletion typically correct the metabolic abnormalities. In patients with minimal alkalosis histamine-2 blockers and anticholinergic therapy can be initiated. In severe cases of alkalosis that are not responsive to more standard therapy omeprazole can be used to inhibit K^+/H^+ ion exchange.[21]

A potentially painful complication of gastrocystoplasty is hematuria-dysuria syndrome, a complex that includes dysuria, genital skin irritation and excoriation, bladder spasms, suprapubic and/or urethral pain and gross hematuria. The etiology is likely related to chemical irritation of the urothelium when exposed to gastric acid. Treatment includes increased fluid intake, correction of potassium abnormalities, use of histamine-2 blockers, anticholinergic agents and omeprazole, and at times removal of the gastric segment.[22] This clinical entity can be a particular problem in the sensate patient and careful consideration should be taken prior to performing gastrocystoplasty.

JEJUNOCYSTOPLASTY

Jejunum should not be used for enterocystoplasty because of the almost uniform metabolic cost. Electrolyte abnormalities are the increased secretion of sodium and chloride with increased reabsorption of potassium and hydrogen ions, resulting in hyponatremic, hypochloremic, hyperkalemic, azotemic metabolic acidosis. The excess loss of sodium chloride causes osmotic diuresis, resulting in severe water loss and dehydration, and stimulation of the renin-angiotensin-aldosterone system. Aldosterone may also increase secondary to hyperkalemia.[23] The high levels of renin and aldosterone facilitate renal sodium reabsorption and potassium loss, producing urine low in sodium and high in potassium. This establishes a favorable concentration gradient for sodium loss and potassium reabsorption by the jejunum, thus, perpetuating electrolyte abnormalities. Hyperalimentation exacerbates the jejunal syndrome, although the mechanism is unclear.[24] Clinically these serum electrolyte abnormalities result in lethargy, nausea, vomiting, dehydration, muscular weakness and fever. Jejunal syndrome is more commonly seen when the proximal jejunum is used[25] and the incidence is affected in a direct relationship with the length of jejunal segment used.[26]

RENAL DETERIORATION

The preservation of renal function is an important goal of therapy in patients undergoing enterocystoplasty and main-