

Fiberoptic Intubation And Monitoring Of Somatosensory Evoked Potentials In Children With Mucopolysaccharidoses

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Abstract

The management of children with mucopolysaccharidosis (MPS) is a challenge for the anesthesiologist, primarily because of difficult airways. Furthermore, certain types of MPS (I, VI, VII) are at risk for cervical spine compression. Therefore, we recommend monitoring of somatosensory potentials during intubation and during positioning of these patient for surgery. Two case reports are presented to demonstrate the perioperative management.

Water intoxication (restlessness, frothing, retching, tremor and twitching of muscles) was first described by Wier et al. in 1922 (1). In 1998 we are still diagnosing and treating iatrogenic water intoxication described as TURP Syndrome. The presented case describes a typical TURP syndrome, which was diagnosed early, treated aggressively, and which led to a good outcome for the patient.

Transurethral resection of the prostate consists of removal of prostatic tissue by means of electro-cautery. The electro-cautery wire loop is positioned in the patient's urethra through a special sheath. The surgical field is visualized through a scope. Irrigation solution is used to distend the bladder, clear the surgical site, and remove blood and resected tissue. Normal saline cannot be used as irrigation solution because the dissemination of the electric current would be dangerous to both, surgeon and patient. A variety of irrigation solutions have been used in the past.

- Distilled water provides excellent optical view but causes intravascular hemolysis due to low serum osmolarity. Subsequent hemoglobin precipitation in the renal tubulus causes acute renal failure.
- Glycine solution has a osmolarity of 200 mOsm/L. Glycine is metabolized in the liver to ammonia and may lead to visual impairment. High ammonia levels may result in neurologic disturbance.

- Mannitol solution is the only isoosmolar irrigant (275 mOsm/L). It is not metabolized and excreted purely by the kidneys. Absorption of larger amounts of mannitol will shift fluids into the vascular compartment and lead to quick fluid overload, pulmonary edema, and cardiac failure.
- Sorbitol solutions are metabolized to CO₂ (70%) and dextrose (30%). Sorbitol is a non-toxic isomer of mannitol. Absorption of large amounts of sorbitol may lead in addition of the complications of fluid overload to hyperglycemia and hypercarbia.
- Cytal solution is a combination of mannitol and sorbitol. Its osmolarity is 178 mOsm/L.

80 min into the case the patient was noted to be markedly hypothermic. This may reflect the large amount of cold solute absorbed by the patient and confirmed by the Na measurement of 109 mEq/L. A decrease in serum Na of 20-30 mEq/L (preoperative Na was 138 mEq/L) implies absorption of 3-4 liters of fluid (dilutional hyponatremia) (2). All other vital signs remained stable. Another clue to a hypervolemic state might be hypertension and reflex bradycardia. Several factors contribute to the rapid volume expansion, namely the intravesicular pressure (governed by the height of the irrigation bag above the prostatic sinuses),

the number of prostatic sinuses opened and maybe most important the duration of the surgical procedure. Surgery was terminated at this point and the patient was transferred intubated and sedated to the ICU.

{image:1}

Slightly increased pulmonary vasculature suggesting fluid overload.

{image:2}

Sinus bradycardia of 52 bpm without QT- or ST- abnormalities

The patient was still intubated and sedated. Therefore, we were unable to assess his neurological function, but we can assume that secondary to his profound hyponatremia, hypoosmolality and high ammonia levels he would have had some neurological abnormality. Some anesthesiologists and urologists would prefer regional (epidural or spinal anesthesia) compared to general anesthesia in order to early recognize mental changes in their awake patients. Neurologic changes (confusion, agitation, or other mental changes) are among the first clinical signs of water intoxication. Early recognition and early treatment of a TURP syndrome is an extremely important factor in its therapy. Neurological symptoms vary through a spectrum from mild confusion and encephalopathy through to seizures and coma. The neurological effects may result from cerebral edema associated with acute hypotonicity or from toxicity of glycine. If glycine concentrations are 30 times the normal value, visual impairment may occur (2). Visual impairment also ranges across a spectrum from blurred vision through blindness, as glycine is a major inhibitory neurotransmitter in the retina (3). Our patient did exhibit some confusion and agitation after awakening. This resolved completely within 48 hours. No visual impairment was noted.

The patient was extubated after 24 hours. The vital signs were stable, the lung clear to bilateral auscultation, and the chest x ray negative for pulmonary edema.

{image:3}

Normal Chest Xray.

The patient's severe hyponatremia was initially aggressively treated with 3% saline. This is a somewhat controversial area. The most feared complication of rapid correction of hyponatremia is the "osmotic demyelination syndrome" (3).

Demyelinating lesions have not been reported after correction of acute hyponatremia in TURP patients, though recent animal studies have shown that rapid Na correction can cause neurological disease (2). Therefore, serum sodium and osmolality should be monitored and corrected only until symptoms subside, then correction should be continued at a Na correction <1.5 mEq/L/Hr. It should be noted that hyponatremia occurs not because of loss of sodium but because of gain of free water. Excessive administration of hypotonic saline results in additional fluid overload (4). NaCl 3 % at a rate of 100 cc/ h was started in the OR just prior to the transfer to the SICU. In addition, 20 mg of furosemide i.v. were administered. Despite this treatment, the sodium level 20 minutes later was 107 mEq/L. Blood samples were sent every hour for the next 6 hours. 1 hour later NA was 116 mEq/L. The 3% saline infusion was stopped and an infusion of 1/ 2 hypertonic normal saline was started at 50cc/hr. Within 24 hours, sodium levels stabilized at 127 mEq/L. After 48 hours, the sodium level reached 132 mEq/L. The patient was discharged from the SICU with blood values within normal limits: NA 134 mEq/L, K 3.5 mEq/L, CL 105 mEq/L, Gluc 99 mg/dl, Bun 15 mg/dl, Creatinine 0.6 mg/dl, Ammonia 24 mmol/L and Serum Osmolarity of 287.

Our patient did not have an extreme hyperammonemia (normal level is 11 to 35 mmol/L). The increase of serum ammonia during TURP is due to glycine absorption, which undergoes oxidative deamination, leading to the formation of glyoxylic acid and ammonia. Extreme hyperammonemia (concentrations > 150 mmol/L) can exacerbate the cerebral depression caused by the hyponatremia. L-arginine, acting in the hepatic tissue prevents hepatic release of ammonia and accelerates the conversion of ammonia to urea.

In conclusion, hypoosmolality and hyponatremia appear to be the principle culprits contributing to the neurological changes seen in TURP-syndrome. Aggressive correction of the above and good supportive care for the renal, cardiac and pulmonary systems contributed to the good clinical outcome for our patient. He was discharged from our ICU without any complications.

QUESTIONS & ANSWERS

1) WHAT IS YOUR DIAGNOSIS ?

TURP-Syndrome

2) WHAT HAPPENED INTRAOPERATIVELY ?

Absorption of 3 to 4 liters of bladder irrigation leading to

hypervolemia, hyponatremia, hypothermia, and hyperammonemia

3) WHAT ARE THE POTENTIAL COMPLICATIONS OF THIS SYNDROME ?

neurologic system:

- confusion to coma
- blurred vision to blindness

cardiovascular system:

- bradycardia or other arrhythmias
- hypertension followed by hypotension
- heart failure

pulmonary system:

- pulmonary edema
- cyanosis, hypoxemia

hematologic system:

- dilutional anemia
- hyperkalemia (hemolysis)
- coagulopathy, bleeding
- high glycine and ammonia levels

4) HOW WOULD YOU TREAT THIS PHENOMENON ?

- Treatment of TURP syndrome should start without delay.
- The surgeon should be informed immediately about the possibility of a TURP syndrome and surgery should be stopped as quick as possible.
- Send blood for measurements of electrolytes, blood gas analysis, hemoglobin and hematocrit, coagulation profile, glucose and ammonia level.
- Switch irrigation to warm normal saline for continuous bladder irrigation in order to avoid further hypothermia and hemodilution with hypoosmolar solutions.

- Continue ventilation or consider intubation (in awake, non-intubated patients) in cases will pulmonary edema and respiratory insufficiency. Administer furosemide. Anti-Trendelenburg positioning if hemodynamically tolerated.
- Monitor for hyponatremia and treat only if clinical symptoms are obvious or very likely. Treatment with hypotonic saline is somewhat controversial as explained above in the case report. Monitor sodium levels frequently during replacement therapy. Avoid quick correction of hyponatremia. Administer maximal 50 to 100cc/hr of 3% saline until clinical signs of hyponatremia disappear. Continue then with diuretic therapy and infusion of normal saline (0.9%).
- Monitor for hyperkalemia due to hemolysis during hypoosmolar dilution and for hypokalemia during treatment with diuretics. Correct potassium imbalances.
- Monitor for coagulopathy and treat disturbances accordingly.
- Consider placement of an arterial catheter, central venous catheter/pulmonary artery catheter for better control of hemodynamics and fluid therapy.
- Treat complications such as seizures or arrhythmias while correcting their causes (hyponatremia, hyperkalemia, hypokalemia, hypoxia, hypothermia).

5) HOW CAN IT BE AVOIDED ?

- limit duration of surgery
- minimize exposure to open venous sinuses by careful surgical resection
- limit the position of the irrigation bag to max. 60 cm above the surgical field in order to minimize hydrostatic pressure of the fluid
- limit extent of bladder distension by frequent drainage of the bladder in order to avoid increased absorption through open venous sinuses
- maintain adequate blood pressure and therefore normal periprostatic venous pressure in order to

avoid increased absorption through open venous sinuses

- check neuro-status (in patients with regional anesthesia), temperature, and laboratory measurements frequently

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