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11 November 08

12 December 08

July: Question 1

You are called to the operating room for an emergency cesarean section of a mother who just had a seizure. The obstetrician hands you a full-term hypotonic infant making only weak respiratory efforts. After an appropriate resuscitation, the infant is pink with a normal heart rate and perfusion, but still somewhat hypotonic. Over the next few hours, the weakness is accompanied by poor feeding, apnea, decreased bowel sounds, and no meconium production. Skin turgor, urine output, and serum glucose concentration are normal. Among other diagnoses, you suspect an electrolyte abnormality.

Of the following, the MOST appropriate treatment for this infant is:

1	calcium, insulin, and glucose
2	extra fluids, calcium, and loop diuretics
3	extra free water
4	normal saline, loop diuretics, hydrocortisone
5	phenylbutyrate and benzoate

You selected 3, the correct answer is 2.

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The most likely electrolyte abnormality in this infant is hypermagnesemia. The treatment most likely to benefit a symptomatic child with hypermagnesemia includes fluids, calcium, and loop diuretics. The other treatment choices are important for cases involving hyperkalemia, hypernatremia, hypercalcemia, or hyperammonemia.

Magnesium ion passes freely across the placenta. Equilibration of maternal and fetal serum concentrations occurs with a 2-hour delay. The normal serum magnesium concentration is between 1.6 mEq/L (1.6 mmol/L) and 2.1 mEq/L (2.1 mmol/L). The most common cause of hypermagnesemia in the newborn is maternal administration of magnesium for preeclampsia, or eclampsia as in this vignette. Other causes of hypermagnesemia include renal failure, rhabdomyolysis, hypothyroidism, and excess magnesium administered by antacids, laxatives, or parenteral nutrition.

Signs and symptoms of hypermagnesemia vary by serum magnesium concentration, and neonates often are affected at lower concentrations than children or adults (Table).

Table

Table. Selected Signs and Symptoms of Hypermagnesemia	
Serum Concentration, mEq/L	**Signs and Symptoms**
3-8	Sedation
	Facial flushing
	Hypotension
	Electrocardiographic changes
5-15	Respiratory depression
	Apnea
	Ileus
	Loss of deep-tendon reflexes
	Weakness
	Coma
20-30	Cardiac arrest

Serum concentrations correlate only approximately with tissue levels and clinical signs. Signs in the neonate can last from hours to days. The elimination half-life of magnesium in premature infants with adequate urine output may be as long as 40 hours. Changes on the electrocardiogram may include sinus bradycardia; prolonged PR, QRS, and QT intervals; atrial fibrillation; heart block; and asystole. Signs and symptoms can be potentiated by the addition of weak neuromuscular blocking agents, such as gentamicin. Death has been reported in some cases, mainly from cardiac arrhythmias.

Treatment of hypermagnesemia in a neonate with good urine output requires supportive care, as well as the removal of any intravenous source of the magnesium. Severe symptomatic cases, especially those with cardiac arrhythmias, may benefit from supplemental fluid administration and loop diuretics to promote magnesium excretion. Intravenous calcium acts as a temporary magnesium antagonist and may reverse arrhythmias and electrocardiographic abnormalities. Exchange transfusion and dialysis have been used successfully in severe cases.

Calcium, insulin, and glucose are used to treat severe hyperkalemia. Bicarbonate and cation exchange resins have also been used. Weakness, ileus, and apnea, as in this vignette, are not common signs of hyperkalemia.

Extra free water in the total daily fluids, as the main treatment mode, is important in the treatment of hypernatremia. Hypernatremic neonates are often hypovolemic, and so may also require volume expanders such as normal saline. Calcium and diuretics, potentially useful for the infant in this vignette with hypermagnesemia, are not ordinarily part of the treatment of a neonate with hypernatremia. Clinical signs of hypernatremia may include irritability and hyperpnea, not present in this vignette.

Normal saline, loop diuretics, and hydrocortisone are part of the treatment of hypercalcemia. Hypercalcemia may present with poor feeding, lethargy, and constipation, as in this vignette. It is unlikely to present in the delivery room, and even less likely than hypermagnesemia, given the probable treatment of the mother with magnesium after her seizure.

Benzoate and phenylbutyrate are useful agents for the treatment of hyperammonemia. They provide alternative pathways for the excretion of waste nitrogen. Benzoate is transaminated from glycine to form hippuric acid, eliminating one nitrogen in the urine. Phenylbutyrate causes one glutamine to form phenylacetyl-glutamine, eliminating two nitrogens. Although transient hyperammonemia may manifest as hypotonia on the first day after birth, as in this vignette, it is unlikely to manifest in the delivery room.

References:

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American Board of Pediatrics Content Specification(s):

Understand the etiology, clinical manifestations, and approach to therapy of hypermagnesemia

Recognize and diagnose the metabolic disorders that lead to coma

Know how to recognize inadequate or excessive water intake by analyzing water intake, urine output, weight change, and serum sodium concentration

Understand the clinical manifestations, laboratory features, and treatment of disorders in the metabolism of the urea cycle

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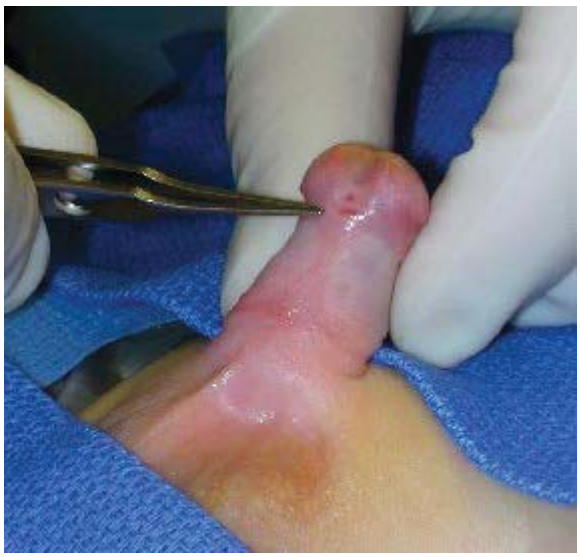
July: Question 9

During rounds with the residents in the newborn nursery, you are asked to see a full-term infant born to a 25-year-old white mother. The mother's pregnancy was uncomplicated. Because of previous pregnancy losses, chorionic villus karyotype was performed and parents were told it was normal. Prenatal ultrasonography performed at 18 weeks' gestation was normal, but parents did not want to know the predicted fetal sex from either study. The infant's abnormal physical finding is seen in the Figure. The chorionic villous sampling result was 46,XY, according to the prenatal records.

Figure



Of the following, the statement that MOST accurately describes details about the evaluation and management of the infant is that:

1	circumcision is recommended in the neonatal period
2	cryptorchidism increases risk of intersexuality
3	meatal obstruction is common
4	renal ultrasound should be obtained
5	surgical repair is usually done by 2 months of age

You selected 2, the correct answer is 2.

The infant in this vignette has penile hypospadias. Hypospadias is a common congenital anomaly of the penis in which the urethra opens proximal to its normal position at the tip of the

glans. Hypospadias, following cryptorchidism, is the second most common genital abnormality in male infants, occurring with an incidence of 0.3% to 0.8% of live births. The incidence of hypospadias has doubled since the 1960s for unknown reasons.

The anatomic location of approximately 87% of hypospadias anomalies are glandular or coronal, 10% are penile, as in the neonate in the vignette, and 3% are penoscrotal or perineal. The anatomy of a penis with hypospadias is similar to that of a normal penis except on the ventral aspect where the foreskin and urethral spongiosum are absent.

Formation of the male external genitalia is a complex process involving genetic programming, cell differentiation, hormonal signaling, enzyme activity, and tissue remodeling. Hypospadias occurs because of an arrest of urethral formation. The urethral opening can be found anywhere along the ventral midline from the perineum to the glans depending on the time during embryogenesis when fusion of the urethral folds ceases. The exact cause of hypospadias is still not known in most cases. In fewer than 5% of patients, hypospadias can be attributed to defects in androgen metabolism (5-alpha-reductase type II deficiency), androgen receptors, or known genetic factors.

Hypospadias is usually identified during the initial examination. The penile raphe is displaced from the midline and the glans tilts downward (chordee). The meatus may be the size of a pinhole but is usually not obstructed. Micturition from the ventrally placed meatus confirms the diagnosis.

Anomalies that may accompany hypospadias include meatal stenosis, hydrocele, inguinal hernia, and cryptorchidism. Evaluation of a newborn with hypospadias may include the following:

- a family history of hypospadias, endocrine, or intersex problems
- a history of possible maternal progestin or estrogen exposure
- examination to evaluate the hypospadias (urethral meatus location, chordee, scrotal folds, phallus length, presence of gonads)
- identification of other congenital abnormalities
- radiographic studies of the kidneys and pelvis if the hypospadias appears to be part of a malformation syndrome

A male infant without cryptorchidism, as in this vignette, who has an isolated urethral opening on the glans or shaft of a normal-sized phallus rarely has a dilemma of gender identity; evaluation for endocrinopathies or intersex disorders are generally not required. Cryptorchidism is defined by the failure of one or both testes to descend completely into the scrotum (at least 4 cm below the pubic crest in a term infant weighing more than 2.5 kg).

Disorders resulting in ambiguous genitalia, such as congenital adrenal hyperplasia in a virilized female, must be considered if the gonads are not palpable and especially if the defect is low on the shaft or scrotum. Congenital adrenal hyperplasia can yield marked virilization making chromosomal diagnosis essential in all cases of ambiguity with cryptorchidism. If the gonads are not palpable in a male (46,XY) neonate with hypospadias, risk of intersexuality approaches 50%. The likelihood of intersexuality is also increased among male neonates whose meatus is positioned in the scrotum or perineum.

Because cryptorchidism may result from insufficient androgen, it is not surprising that hypospadias and intersex disorders may coexist. Approximately 8% of boys with hypospadias have at least one undescended testicle with the incidence varying with the severity of hypospadias. Only 5% of genetically male neonates with distal hypospadias, as in this vignette, have cryptorchidism, compared with 32% of neonates with proximal lesions.

Hypospadias, often associated with cryptorchidism, occurs in a number of syndromes. The neonate in the vignette has mild hypospadias, defined as glandular or penile, and no other dysmorphic features. Isolated penile hypospadias without other genital abnormalities or dysmorphic features is unlikely to be associated with a chromosomal abnormality.

In the past, boys with hypospadias routinely underwent intravenous pyelography, voiding cystourethrography, and renal ultrasonography as part of their evaluation. However, even with severe hypospadias the arrest in development is after the eighth week of gestation, when the urethral bud joins the metanephros. The likelihood of detecting an upper urinary tract anomaly is low, with clinically significant abnormalities found in fewer than 5% of neonates with hypospadias. Radiographic studies may be helpful in boys who develop a urinary tract infection or those whose anomaly is part of a malformation syndrome.

The only treatment for hypospadias is surgical repair of the defect. The goal of surgical correction is to create a penis with normal function and appearance. Because the spectrum of severity is wide, ranging from glandular to perineal, and the shaft may be straight or significantly curved, it is not surprising that more than 300 operative techniques for hypospadias repair have been described. Depending on the severity of the lesion, surgical techniques can be applied sequentially or in combinations beginning at 6 months in an otherwise healthy newborn. In severe cases requiring a two-stage procedure, the second stage is performed at least 6 months after the initial repair. Neonatal circumcision is contraindicated in such cases because foreskin tissue is often used in the repair process.

References:

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Palmert MR, Dahms WT. Abnormalities of sexual differentiation. In: Martin RJ, Fanaroff AA, Walsh MC, eds. *Fanaroff and Martin's Neonatal-Perinatal Medicine Diseases of the Fetus and Infant*. 8th ed. Philadelphia Pa: Mosby Elsevier; 2006:1550-1596

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American Board of Pediatrics Content Specification(s):

Know how to evaluate and manage an infant with hypospadias and epispadias

Know how to evaluate and manage an infant with cryptorchidism

Know the etiology and diagnosis of an infant with ambiguous genitalia, including congenital adrenal hyperplasia

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