Chapter 27
Genitourinary Disorders

Joseph P. Cravero

A baby in the first day of life presents for closure of an exstrophy of the bladder. The product of 36 weeks gestation, he is 2.3 kg, with a preoperative hematocrit of 56 %, BP 65/35 mmHg, HR 130 bpm, and RR 24/min.

J.P. Cravero, MD, FAAP
Department of Anesthesiology, Perioperative, and Pain Medicine, Boston Children's Hospital, Boston, MA, USA

Harvard Medical School, Boston, MA, USA
e-mail: joseph.cravero@childrens.harvard.edu

R.S. Holzman et al. (eds.), *Pediatric Anesthesiology Review*,
DOI 10.1007/978-3-319-48448-8_27

Preoperative Evaluation

Questions

1. Is this baby premature? How can you differentiate premature from small for gestational age (SGA) babies? What difference would it make in your anesthetic technique? What problems would you expect related to prematurity? Should a regional anesthetic be utilized? Narcotics? Why/why not? Would you hope to extubate this baby at the end of surgery?

Preoperative Evaluation

Answers

1. Yes, because the infant was born before 37 weeks gestation. Infants born between 36 and 37 weeks gestational age are categorized as borderline premature. Those born between 31 and 36 weeks GA are considered moderately premature and those born between 24 and 30 weeks gestation are considered severely premature. Small gestational age babies weigh less than 2.5 at birth. The more premature the infants are, the greater the risk for perioperative complications. Premature babies are born with structurally and physiologically underdeveloped vital organs. They are unable to maintain body temperature due to immature thermal regulation. Hyperthermia metabolic rate linearly between 36 and 28°C, therefore increasing oxygen consumption, which can lead to hypoxemia, acidosis, apnea, and respiratory depression. Premature infants tend to lose body heat at a faster rate than term or older infants because of a higher body surface/volume ratio and lack of brown fat. Heat stress is equally detrimental because premature infants are unable to sweat (dissipate heat by evaporative heat loss), and body heating causes dilation of peripheral vessels. During anesthesia the infant's body and head should be covered with plastic or cotton wrap to decrease heat and water loss [1, 2].

   Infants are unable to sustain ventilation due to poorly developed ventilatory centers in the brainstem and inefficient respiratory mechanics. Premature infants are also at risk for respiratory distress syndrome due to impaired amounts or lack of surfactant. They may also develop intraventricular hemorrhage from rapid changes in blood pressure or cerebral ischemia from hypoperfusion due to impaired cerebral autoregulation. In addition, this population is at risk for left to right shunting via the ductus arteriosus soon after birth (within 3–5 days after birth). Premature infants born before 34 weeks gestational age have a decreased glomerular filtration rate (GFR). Even term neonates have only 40 % of an adult's GFR at birth. In addition, there is decreased tubular reabsorption capacity and a relative inability to absorb water, salts, glucose, protein, phosphate, and bicarbonate. Hyperglycemia and glycosuria can act as an osmotic diuretic and cause obligatory sodium as well as free water loss. Hepatic catalyzing enzymes are less active in premature infants. Oxidizing, reducing, and hydrolyzing enzymes are relatively inactive. Conjugation enzymes (conjugation with acetate, glycine, sulfate, and glucuronic acid) are also less active except for sulfonation. Therefore, the metabolism of various drugs, particularly opioids, can be impaired. These enzymes mature between 6 and 12 months of age, to adult capacity. A regional anesthetic should be used whenever feasible. Opiates could be used with caution, in reduced doses, and the infant's respiratory status should be closely monitored. I would hope to extubate if successful epidural analgesia is provided and minimal opiates are administered intraoperatively. Prior to placement of a caudal or epidural, radiological images of the spine (which almost certainly would already have been part of this child's work-up) should be evaluated to ensure the anatomy is normal.

2. Is it common for bladder exstrophy to occur in males? What do we call bladder exstrophy in a female? Why does this happen? Are there any future problems the patient can expect? Is early closure better than later closure? Why/why not? Is it likely that there is more surgery in the future for this baby? What type? Why?

Intraoperative Course

Questions

1. Does this baby need an arterial line? Why/why not? Should a central line be placed? Where would you place the IVs? Why? Can you only get an IV in the foot? What next? If the case will take 8 h, do you need to obtain surveillance blood gases? Why/why not? Would you treat if the pH were 7.34? 7.22? 7.14? Why?

2. Can you do this case with an epidural or caudal catheter and sedation? Would you choose to do so? Why/why not? A general anesthetic with an endotracheal tube is chosen. Would you place an oral or a nasal tube? Why? Should narcotics be avoided? Which would you choose? Why/why not? What about muscle relaxants? Surgeon wants you to avoid them. (Why do you think?) Is nitrous oxide contraindicated? Relatively contraindicated? What problems might you expect? When would you expect them?

2. Yes; the male/female ratio is 2:1. In a female, bladder exstrophy is known as a cloaca. At 5–6 weeks of gestation, the cloacal membrane prevents the normal migration of mesoderm (originator of anterior abdominal muscles and pelvic bones) of the infraumbilical area resulting in failure of fusion of the rectus muscles and the pubic symphysis; the urethra fails to close dorsally (epispadias), and the anterior wall of the bladder wall may remain open. The urinary tract is everted exteriorly. Future problems include incontinence and sexual dysfunction. Early closure (within 24–48 h) of the bladder and abdomen may allow an optimal anatomical and functional outcome. This child will likely require many further reconstructive surgeries to correct epispadias at age 2–3 years and urinary continence (the bladder neck) by age 4–5 years. Other possible procedures include bladder augmentation if the bladder is of small capacity, ureteral reimplantation for ureteral reflux, and creation of a continent urinary (e.g., Mitrofanoff) stoma [3].

Intraoperative Course

Answers

1. An arterial line would be very helpful as it would enable surveillance blood gases and monitoring of accurate and continuous blood pressure (which are critical in cases with significant blood loss and large fluid shifts at this age). A central line is not necessary unless a prolonged postoperative course is expected along with difficulty in intravenous access. In that case, it may be wise to place a percutaneously inserted central catheter (PICC) line as part of the procedure. IVs should be placed in the upper extremities if possible because the lower extremities are usually prepared and draped within the surgical field and because of potential for loss of infusate from iliac veins within the surgical field. Surveillance blood gases are a good idea for a patient this age undergoing prolonged surgery to monitor accurate and continuous blood pressure, blood loss, and large fluid shifts. I would be cautious with a pH of around 7.25 and treat a pH less than 7.22 because it is associated with deleterious acidosis and impaired cardiovascular performance.

2. No, this case cannot be done with a regional technique alone. This is a prolonged procedure that would require a dense block sustained for a long period of time. The use of an adequate surgical concentration of local anesthetics in premature infants could result in systemic toxicity due to impaired hepatic elimination of amide and ester local anesthetics resulting in prolonged elimination half-lives. Moreover, sedation would be required in addition to regional anesthesia and could result in hypoventilation, periodic breathing, and apnea due to immaturity of the respiratory centers. The amount of sedation required, even with an optimal block, would result in alveolar collapse, shunting, and respiratory embarrassment with a small infant positioned very far away from the anesthesiologist. A nasal or oral endotracheal tube would be acceptable. I would use a nasal tube

3. The surgeon has turned the baby prone and is in the process of doing bilateral iliac osteotomies. The saturation drops to 94 % and the end-tidal CO_2 has disappeared from the screen. What do you think is going on? The blood pressure cuff is not reading but recycling. The electrocardiogram (ECG) heart rate is 110? Then it becomes 80? What is Durant's maneuver? Should the baby be placed in the left or right lateral decubitus position?

Postoperative Course

Questions

1. Should this patient be extubated? What criteria would you use? The patient is vigorous, and you extubate, but saturation is 92 % on supplemental $oxygen_2$ by face shield? Your next move? Face mask fails to improve saturation; would you be happy with 93 %? What if it was 91 %? Should patient be reintubated? For a respiratory rate of 24/min? 34/min? 54/min? What could be going on to account for the findings? How would you manage V/Q mismatch at this time?

2. How would you manage a continuous morphine infusion for pain relief? What about an epidural? How would you constitute the epidural solution? With fentanyl or Dilaudid? Why? A colleague stops by as you are putting in the epidural needle and says that he always gets a spine film in babies with bladder exstrophy because he is worried about *spinal anomalies that would complicate placement and impair appropriate spread of the local anesthetic?* Is he right? What difference would it make in your anesthetic management? Could you place a continuous caudal catheter instead? Or would you just stay away from the back?

because it would be more secure intraoperatively and comfortable for the infant if he requires postoperative ventilatory support. Opiates should not be avoided. I would choose fentanyl, because the mixed enzyme oxidase enzymes are adequately mature to metabolize fentanyl effectively. The mixed oxidase enzymes that metabolize fentanyl are more active in early infancy. Muscle relaxants should be avoided if the surgeon plans to stimulate and assess sphincter function or identify major nerves. While nitrous oxide is not absolutely contraindicated, it is relatively contraindicated due to the fact that it can accumulate in the bowel causing distension, or aggravate air embolism, should it occur.

3. Possible air embolism, occlusion or dislodgment of the endotracheal tube, or severe hypotension; most likely this is an air embolism. Durant's maneuver involves positioning the patient in steep head down and left lateral decubitus position. Left lateral decubitus allows the buoyant foam (blood/air mixture) to remain in the right ventricle and prevent it from occluding the pulmonary arteries.

Postoperative Course

Answers

1. It is perfectly reasonable to plan for extubation, assuming some specific criteria are met: if the infant is awake, exhibits adequate strength (flexing at the hips for 5 s, tight fists, strong bite, furrowing his eyebrows as a sign of attention), the muscle relaxant (if used during surgery) is reversed, with a return of train of four and no fade at 50 Hz (for 5 s tetanus), is breathing regularly, and has an empty stomach. If oxygenation were poor after extubation, I would provide 100 % oxygen with a face mask. I would not be happy with a saturation of 93 % or 91 % and would reintubate if this persisted. Hypoxemia and hypercarbia are likely due to ventilation/perfusion mismatch which should be improved with positive end expiratory pressure (PEEP) and appropriate peak ventilator pressures. I would plan controlled ventilation after reintubation of the trachea.

2. I would start the morphine infusion at 15 mcg/kg/h after intubation or 10 mcg/kg in a non-intubated spontaneously breathing infant. Placement of a caudal-to-thoracic or lumbar catheter and infusion of local anesthetics is a safe alternative to IV opioids. Appropriate placement could be confirmed by ultrasound, epidurogram, or electrical stimulation. A solution of chloroprocaine 1.5 % with or without fentanyl (1–2 mcg/cc) is safe in this age group and provides appropriate analgesia. If the trachea is extubated, it may be prudent to avoid neuraxial opioids to avoid the possibility of opioid-induced periodic breathing and/or apnea. My colleague is probably worried about possible associated vertebral anomalies and is correct about his concern. Identification of normal anatomy is a prerequisite

Additional Questions

Questions

1. What is the difference between a neuroblastoma and nephroblastoma (Wilms' tumor)? Any other syndromes associated with neuroblastoma? Can both be endocrinologically active? What is the difference between a ganglioneuroma and a neuroblastoma? What makes a neonatal Wilms' tumor (congenital mesoblastic nephroma) different from a regular Wilms' tumor?

2. What is the natural history of "infantile" polycystic kidney disease? How does it affect anesthetic management? What are the nonrenal considerations for perinatal form?

3. You wish to perform a peripheral nerve block for hypospadias repair. Which nerves do you wish to block and what anatomical structures will your needle pass through on the way to those nerves?

for insertion of neuraxial needles and catheters. A continuous caudal catheter could be placed if there is no associated sacrococcygeal agenesis or anomaly. Placement of an epidural indwelling catheter is possible if the spine X-ray reveals normal anatomy at the site of the insertion, and ultrasound, CT, or MRI of the spine reveals no cord tethering [4].

Additional Questions

Answers

1. Neuroblastoma is a neural crest malignancy that arises from primitive blast cells of the postganglionic sympathetic chain and adrenal glands. Nephroblastoma (Wilms' tumor) is a malignancy that arises from abnormal metanephric differentiation of the renal blastema (undifferentiated renal cells). Neuroblastoma can be associated with pheochromocytoma and neurofibromatosis type 1 (NF-1, von Recklinghausen disease). Neuroblastoma and nephroblastoma can both be endocrinologically active; 75 % of neuroblastomas may secrete catecholamines. Ganglioneuroma is a benign tumor arising from well-differentiated and mature sympathetic ganglia. Neonatal Wilms' tumor is a benign nephroma arising from metanephric blastema or secondary mesenchyme, whereas regular Wilms' tumor (nephroblastoma) is a malignant tumor of the undifferentiated metanephric blastema.

2. Infantile polycystic kidney disease varies in severity. When oligohydramnios presents early in pregnancy, the outlook is extremely poor due to fetal pulmonary hypoplasia in addition to renal insufficiency. The condition sometimes presents later in infancy with reduced renal function. It may not become symptomatic until adolescence, when it represents a milder expression of the disease. The extent of pulmonary hypoplasia determines the difficulty of ventilation. The degree of renal impairment determines the clearance of anesthetic agents. The obstetrical team may consider cesarean section, particularly in bilateral polycystic kidney disease, because of the large body size and the risk of renal rupture during vaginal delivery. The newborn may require control of ventilation and treatment of high blood pressure.

3. Penile tissues are innervated by the dorsal penile nerve (S 2, 3, 4) and the perineal cutaneous nerve (branch of the pudendal nerve) at the root of the penis. The dorsal penile nerve is best reached at the base of penile shaft from the dorsal surface. The needle has to traverse the skin, subcutaneous tissue, and Buck's fascia. The fascial planes can be visualized with ultrasound, which has been shown to improve success of this block [5]. A subcutaneous ring block of the penis will include a portion of the perineal nerve, but this will depend on the proximal vs. distal location of the hypospadias.

References

1. Williams A, George PE, Dua V. Anesthetic considerations in a preterm: extremely low birth weight neonate posted for exploratory laparotomy. Anesth Essays Res. 2012;6(1):81–3.
2. Taneja B, Srivastava V, Saxena KN. Physiological and anaesthetic considerations for the preterm neonate undergoing surgery. J Neonatal Surg. 2012;1(1):14.
3. Kost-Byerly S, Jackson EV, Yaster M, Kozlowski LJ, Mathews RI, Gearhart JP. Perioperative anesthetic and analgesic management of newborn bladder exstrophy repair. J Pediatr Urol. 2008;4(4):280–5.
4. Veneziano G, Iliev P, Tripi J, Martin D, Aldrink J, Bhalla T, et al. Continuous chloroprocaine infusion for thoracic and caudal epidurals as a postoperative analgesia modality in neonates, infants, and children. Paediatr Anaesth. 2016;26(1):84–91.
5. Faraoni D, Gilbeau A, Lingier P, Barvais L, Engelman E, Hennart D. Does ultrasound guidance improve the efficacy of dorsal penile nerve block in children? Paediatr Anaesth. 2010;20(10):931–6.