

to facilitate intubation. This failed to produce any evidence of muscle relaxation after five minutes. She was then given 80 mg. of succinylcholine intramuscularly and satisfactory relaxation was obtained. Another intravenous route was established in the left arm, utilizing a 15-gauge Rochester needle. The cutdown catheter was withdrawn approximately 5 cm. before blood could be aspirated freely. It was then secured in this position, and dextrose 5 per cent in water with a 0.1 per cent succinylcholine drip was started. Approximately one hour after anesthesia was induced, ventilatory difficulties became apparent, and expiratory "wheezes" were noted bilaterally on auscultation of the chest. Respiratory excursions of the chest were definitely limited on the right and breath sounds on the right were significantly reduced. The endotracheal tube was removed and re-inserted without any improvement in ventilation. Bronchodilators, aminophylline and Bronkephrine, produced only transient improvement in ventilation. During this period of inadequate ventilation, the pulse rose from 80 to 140 per minute with little change in blood pressure. The patient was obviously cyanotic. The period of ventilatory difficulty lasted throughout the two-hour operative procedure, during which period the patient had received 700 ml. of dextrose 5 per cent in water via the cutdown. A radio-opaque dye was injected into the cutdown catheter and chest films were obtained in the operating room. This revealed the tip of the intravenous catheter in the superior vena cava and opacification of the right hemithorax. Increased density in

the medial aspect of the right lung field was felt to represent atelectasis of a portion of the right lung. Thoracentesis was done on the right, and 350 ml. of blood-tinged fluid was obtained. There was immediate and dramatic improvement in the patient's color and ventilation. Chest films taken two days postoperatively revealed considerable clearing of the appearance of the right lung and some clearing of the previously observed atelectasis. Chest films taken six days postoperatively revealed complete clearing of the pleural effusion and atelectasis.

It is believed that the sequence of events in this case was due to dissection of the polyethylene catheter into the right pleural space and the subsequent infusion of fluids into this space. The problems that occurred may have been anticipated from the initial incomplete response to the drugs administered via the cutdown catheter. This represents a potentially serious complication of venous cutdowns that could very well prove fatal if unrecognized.

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Anesthetic Management of Wiskott-Aldrich Syndrome

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The essential triad for the diagnosis of the Wiskott-Aldrich syndrome is thrombocytopenia, eczema and recurrent infections.^{1,2} The type of infection varies; otitis media,²⁻⁴ pyoderma,^{2,}

^{3,5} septicemia,²⁻⁴ meningitis⁵ and pneumonia.^{5,6} Additional features include bloody diarrhea,² anemia,^{2,3,7} epistaxis,⁸ splenomegaly,^{2,3,7} eosinophilia,^{3,7} increased serum globulin,³ and a fatal outcome. The condition is transmitted via a familial sex linked recessive gene carried by the female and manifested only in male infants, much like hemo-

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Supported by NIH Grant GM-09069-03.

philia. Since the description of the syndrome in 1937, 20 families,¹⁻¹⁷ to the best of our knowledge, have been reported which carry this abnormality. Although this syndrome is uncommon, the anesthesiologist should be familiar with the clinical findings since this patient may present for splenectomy.

CASE REPORT

This 25 month old white male was admitted to Babies Hospital because of fever, vomiting, pallor and jaundice of 24 hours duration. Family history: He was the third child of unrelated parents. His older brother had died at age 3½ of intracranial bleeding. He had suffered from thrombocytopenia, eczema, recurrent otitis and melena. An older sister is healthy. The mother's brother died before she was born of bleeding and thrombocytopenia. Growth and development: The child was born 3 weeks before term and weighed 5 pounds and 3 ounces. Petechiae were noted at birth, but the child was never jaundiced, cyanotic or anemic. A bone marrow biopsy had shown normal megacaryocytes with decreased platelet formation. He bruised easily and had several courses of steroid therapy to control the thrombocytopenia. He walked at 15 months and spoke single words at 2 years of age. When he received live measles vaccine after 2 doses of killed vaccine, he developed a large vesicular eruption resembling a primary smallpox take. Therefore he was not immunized against smallpox.

Review of Systems. Significant findings include dry, scaly skin with eczematoid eruption at age one year, frequent upper respiratory infections with otitis media, two episodes of pneumonia, the most recent one year prior to admission, and frequent dark, tarry stools. There was no history of hepatitis, hematemesis, convulsions or fainting.

Physical Examination. On admission the temperature was 102° F., pulse rate 156, respiratory rate 88, blood pressure 90/65 and the child weighed 10.6 kg. Abnormal physical findings included severe jaundice, a liver edge palpable to the umbilicus and a spleen palpable 7 cm. below the left costal margin. There was ecchymosis over the extremities and forehead.

Laboratory Investigations. On admission the hemoglobin was 2.5 g./100 ml. with hematocrit of 15 per cent. The white cell count was 11,500 with 68 per cent neutrophils, 23 per cent lymphocytes and 10 per cent monocytes. The urea nitrogen was 17 mg., the sodium was 150 mEq./liter, the potassium 4.0 mEq./liter, the chloride 109 mEq./liter, and the carbon dioxide 19 mEq./liter. The serum bilirubin was 10.9 mg./100 ml. and the Coombs test was 4 plus positive. The bone marrow was hypocellular; only one megacaryocyte was seen but there was active granulopoiesis. The spinal fluid was clear with a total protein of 10 mg./100 ml. and a glucose of 76 mg./100 ml. There were no white blood cells and 20 red blood cells per cubic millimeter. The urine was dark with a specific gravity of 1.017, containing a trace of albumin but free of sugar and acetone.

Hospital Course. During the 80 hours preceding operation the patient received sodium bicarbonate, antibiotics, large doses of steroids and ACTH and a total of 2,000 ml. of whole blood. The hematocrit increased to 33 per cent after transfusion. An increase in the serum bilirubin to 90 mg./100 ml., persistent fever, and lack of improvement following steroid therapy prompted an emergency splenectomy in an attempt to control the auto hemolysis.

Anesthetic Management. The child was given 0.3 mg. scopolamine, intramuscularly 45 minutes prior to his arrival in the operating room. Anesthesia was induced with 30 per cent cyclopropane in oxygen followed by endotracheal intubation with a no. 2 portex tube. Anesthesia was maintained with 10-20 per cent cyclopropane in oxygen delivered through an infant circle semiclosed system. Respirations were controlled. Shortly after operation began, succinylcholine (15 mg.) was administered intravenously to relax the abdominal wall. Within 30 seconds the pulse rate fell from 120 per minute to 30 per minute with a simultaneous fall in blood pressure from 100/70 to 75/40. Release of traction and decreasing the concentration of cyclopropane resulted in a return of the pulse rate to 120 per minute within 4 minutes. A few minutes later, in the absence of surgical stimulation, a 10 mg. dose

of succinylcholine again caused a profound bradycardia. This has been reported previously.¹⁸⁻²¹ After the return of spontaneous respirations *d*-tubocurarine (3 mg.) was administered for muscle relaxation. At the end of the 2-hour procedure, respirations were adequate and antagonism of the relaxant was not deemed necessary.

The child developed pneumonia on the fourth postoperative day, a common complication following splenectomy in young children.^{22, 23} He recovered from the pneumonia and is at present doing well.

SUMMARY

Anesthetic experience with a patient with the Wiskott-Aldrich syndrome is presented. A noteworthy complication was serious bradycardia after two doses of succinylcholine during cyclopropane anesthesia.

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