



Pediatric Clips

Child presents with hereditary spherocytosis —
Thomas Curran, MD

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Pediatric Clips from The Children's Medical Center are quick reviews of common pediatric conditions.

The Children's Medical Center is the region's pediatric referral center for a 20-county area. As the only facility in the region with a full-time commitment to pediatrics, Children's offers a wide range of services in general pediatrics as well as in 35 subspecialty areas for infants, children and teens. We welcome your inquiries about services available — call 937-641-3666 or e-mail marketing@childrensdayton.org.



Dayton, Ohio

All kids need special care —
All kids need Children's.

CASE: CHILD WITH HEREDITARY SPHEROCYTOSIS

Emily is an 8-year-old female whose father has hereditary spherocytosis (HS). He had undergone a splenectomy at 7 years of age. Her father has done well since, but has a large left subcostal scar. Emily has not had any episodes of jaundice, but has recently developed intermittent abdominal pain. On physical examination, she appears healthy, but slightly pale, with a palpable spleen. An ultra-

sound examination showed only splenomegaly with no evidence of gallstones. Her hemoglobin is 11.1 gm/dl and her reticulocyte count 7.5%. Her osmotic fragility test was consistent with HS.

Elizabeth received immunizations for pneumococcus and meningococcus and was electively admitted to The Children's Medical Center of Dayton for a laparoscopic

splenectomy. This was performed uneventfully using three port sites, and she went home the following day. She was placed on penicillin prophylaxis. A month later, her hemoglobin was 15.0 gm/dl, and her reticulocyte count was 1.5%. Her parents reported an increased energy level and there have been no more complaints of abdominal pain.

CASE DISCUSSION

Hereditary spherocytosis is a common, autosomal dominant, hemolytic anemia, affecting approximately one in 5,000 persons of Northern European ancestry. It is the most common condition requiring elective splenectomy in the pediatric age group. Most patients will have an affected parent, but sporadic cases do occur. The condition is due to a deficiency in spectrin, a structural protein in the red blood cell membrane. The unstable membrane causes the cells to become sequestered and destroyed in the spleen.

The disease can vary from mild to severe. It is one of the causes of neonatal jaundice and may be confused with ABO incompatibility. During childhood, most patients have mild splenomegaly without other symptoms. Chronic fatigue, malaise and abdominal pains are common complaints. Older children may develop gallbladder symptoms from



pigmented calculi forming due to the chronic hemolysis. Hemolytic or aplastic crises may occur, often associated with infections. Severe episodes may lead to the need for transfusion.

Laboratory studies typically show mild chronic anemia, with hemoglobin between 9-11 gm/dl.

The reticulocyte count is elevated. The bone marrow shows typical erythroid hyperplasia of hemolytic anemia. The osmotic fragility is increased.

Splenectomy is the treatment of choice for HS because it will return the red cell survival to normal and eliminate associated symptoms. Except in especially severe cases, it is delayed until the child is at least 5 or 6 years old, to decrease the risk of post-splenectomy sepsis. Post-splenectomy sepsis, although rare, can have a significant mortality. The asplenic patient is particularly at risk for overwhelming infections from encapsulated organisms, hence, the rationale for immunizations against pneumococcus, meningococcus and haemophilus influenzae B. The immunizations are generally given two to four weeks prior to the procedure, and penicillin prophylaxis is continued afterward. Families are instructed to seek immediate medical attention for fever and to make sure their immunizations

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stay current. Ultrasound examination is recommended preoperatively to screen for gallstones, as concomitant cholecystectomy may be necessary.

Laparoscopy has become the method of choice for elective splenectomy. Several studies have shown that it is now equally as safe as open splenectomy, with no greater complication rate. Laparoscopy offers less pain, better cosmetics and a shortened hospital stay in contrast to the open procedure. As in

this case, many patients go home the following day with small laparoscopic incisions. This is often in stark contrast to the affected parent who carries a large scar and may have spent a week in the hospital for splenectomy.

REFERENCES

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FEATURED SPECIALIST



THOMAS CURRAN, MD, is a pediatric surgeon at Dayton Children's. Dr. Curran is board certified by the American Board of

Surgery in general surgery, pediatric surgery and surgical critical care. He received his medical degree from Keck School of Medicine of the University

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GENERAL PEDIATRIC SURGERY

Pediatric Surgeons of Dayton, Inc. is a private practice located at Dayton Children's. It offers a level II certified pediatric trauma service. The general surgery division at Dayton Children's offers consultation and care for a variety of problems in infants, children and adolescents with emphasis on congenital, neoplastic and traumatic conditions.

CONTACT INFORMATION

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