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Annular Pancreas

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

The pancreas is an organ that sits behind the stomach. It makes chemicals (called enzymes) that help in the digestion of food. It also makes insulin, a chemical (hormone) that controls blood sugar. In most people, the pancreas is a flat organ deep in the belly. It is located in between the stomach and the spine just above the belly button. The right edge of the organ sits next to the small intestine, a portion called the "duodenum".

In annular pancreas, the shape of the pancreas is different. A ring of pancreatic tissue surrounds a portion of the duodenum (Figure 1). This situation can cause narrowing of the duodenum, leading to vomiting, or it can cause no problems at all. This condition does not affect the function of the pancreas as it continues to make enzymes for digestion and insulin.

How does it happen? When a baby is developing inside the mother during pregnancy, the pancreas starts as two parts (called buds). These buds are supposed to come together and fuse to the left of the duodenum. In annular pancreas, the pancreatic buds do not come together, they fuse around the duodenum instead of next to it.

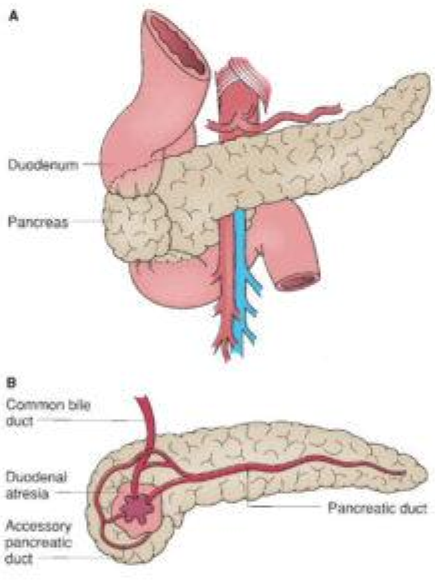

Figure 1: Annular Pancreas around the duodenum

The exact number of people affected is not clear. Most people with this condition have no problems, so they do not seek medical help. It is estimated to occur in 5-15 people per 100,000 (less than 1 percent).

Annular pancreas is sometimes seen associated with other defects that are present at birth (congenital) such as Down's syndrome, problems with the esophagus and duodenum, malrotation and heart defects. In some people, the abnormal shape of the pancreas can lead to narrowing or blockage of the small intestine.

Signs and Symptoms - "What symptoms will my child have?"

Approximately one-third of patients with annular pancreas develop symptoms. When symptoms occur depends upon the severity of the intestinal blockage. Those with severe blockage present as a baby with vomiting, inability to tolerate milk or formula. Sometimes the vomited fluid has bile (color green or yellow).

Later signs/symptoms seen in older children include belly pain, feeling full early during meals, vomiting, not wanting to eat.

If a baby or child has bright green or yellow vomit (mixed with bile), he or she should be seen immediately by a doctor. Sometimes vomiting bile can be because of a very serious condition that may be life threatening (volvulus).

Diagnosis - "What tests are done to find out what my child has?"

Since most people with annular pancreas have no symptoms, there are usually no abnormalities on blood tests or X-rays.

In those patients who have symptoms or blockage of the duodenum because of the annular pancreas:

Prenatal ultrasound may show *maternal polyhydramnios.* Maternal polyhydramnios is when excess fluid is present around the fetus. In addition, there may be a "double bubble sign" that suggests narrowing of the duodenum.
X-rays taken after birth may reveal a "double-bubble sign" or evidence of a duodenal blockage.

Figure 2: Abdominal X-ray demonstrating a double bubble sign.

Sometimes CT scans (Computerized Tomography—multilevel X-rays of the body) or MRIs (Magnetic Resonance Imaging—radiology study that uses magnetic energy to view the inside of the body) may be done to fully assess the anatomy and confirm the diagnosis prior to surgery.

Conditions that resemble this condition include intestinal malrotation, duodenal (intestinal) atresia or abnormal position of a vein that goes to the liver (portal vein).

Treatment - "What will be done to make my child better?"

Surgery is done only for annular pancreas that leads to narrowing of the intestine.

The goal of surgery is to make a way for the food to pass from the stomach into the intestine. Usually, the part of the intestine before the blockage is sewn to the part of the intestine after the blockage. This allows the food to "bypass" the blockage. Removal of the intestine is almost never needed.
Depending on the severity of the blockage, age of the child and other factors, the surgery can be done using a bigger incision (open procedure, laparotomy) or several small incisions (laparoscopic, minimally invasive).

Preoperative preparation: If your child is dehydrated from vomiting, he or she will be given intravenous (IV) fluids. In some cases, the child has malnutrition from vomiting and inability to tolerate food. The doctor will decide whether the child needs intravenous nutrition or tube feedings prior to the operation.

If other anomalies are suspected, then tests may be done prior to surgery to assess for these conditions. For example, an echocardiogram (heart ultrasound) may be ordered if heart defects are suspected.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Postoperative care: Your child will recover in a monitored surgical ward. He or she will not be allowed to eat right away to allow healing of the intestines. They will receive medicines for pain. Possibly intravenous nutrition will be given until feedings are started.

The length of the hospital stay depends upon the child's age, postoperative complications, and severity of the blockage.

Home Care - "What do I need to do once my child goes home?"

Diet: Once the patient starts passing gas and stool and is not vomiting any longer, he or she does not have dietary restrictions and are often allowed to resume a normal diet.

Activity:

- Infants are under no restriction but may not be able to tolerate tummy time for the first few weeks following surgery.
- In older children, recovery depends on the approach. If the laparoscopy was used, the child can get back to normal in about two weeks. If an open or laparotomy approach is used, it will be 4-6 weeks for the incision to completely heal. During this time, children are encouraged to refrain from physical education, contact sports or strenuous activity. The exact instructions postoperatively will be discussed by your surgeon.

Wound care: Specific wound care instructions will be given by your doctor prior to discharge. Scars will soften and fade over time but will grow with the child.

Medicines: Medication for pain such as acetaminophen (Tylenol) or Ibuprofen (Motrin or Advil) or something stronger like a narcotic may be needed to relieve pain for a few days after surgery. Stool softeners and laxatives will encourage regular stooling after surgery, especially if narcotics are still needed for pain.

What to call the doctor for: If the child has a fever greater than 101.5ºF or 38.5ºC that is not relieved by Tylenol; redness or drainage from the incision; or vomiting mandate a call to your doctor or evaluation in the Emergency Department.

Follow-up care: Follow-up appointments for a wound check and symptoms are scheduled for the first two months. Once a full recovery is noted, then only routine follow-up with the pediatrician is needed.

Long Term Outcomes - “Are there future conditions to worry about?”

Long-term results are excellent. Prognosis often depends on other associated abnormalities. Sometimes the duodenum just next to the narrowing can get very dilated and the muscles may not work properly. If a baby or child with surgical repair of a duodenal narrowing associated with annular pancreas has vomiting that continues for weeks, then a doctor should see the patient.

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