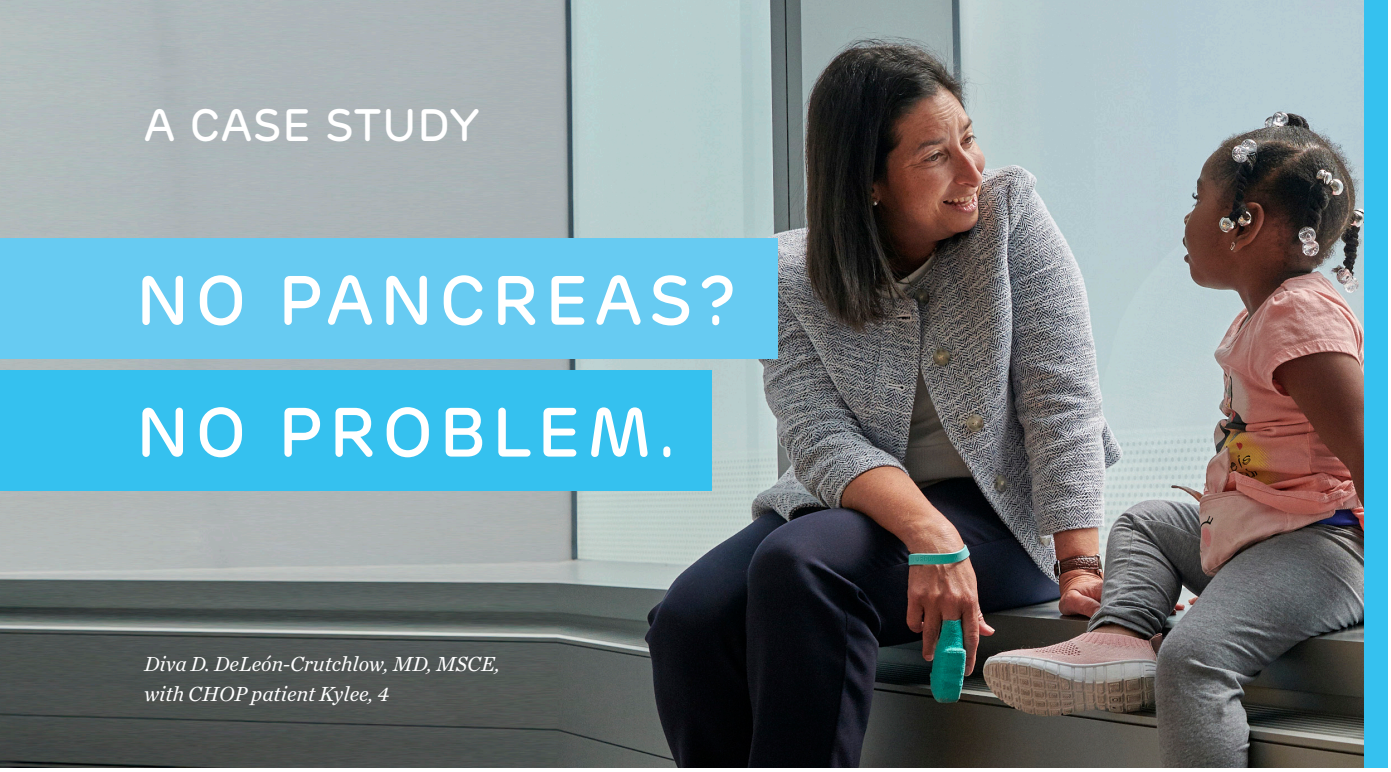
A CASE STUDY

# NO PANCREAS?

# NO PROBLEM.



*Diva D. DeLeón-Crutchlow, MD, MSCE, with CHOP patient Kylee, 4*

## UNIQUE CLINIC AT CHILDREN'S HOSPITAL OF PHILADELPHIA FOLLOWS POST-PANCREATECTOMY PATIENTS

*By Diva D. De León-Crutchlow, MD, MSCE, and Asim Maqbool, MD*

In 2004, R.A. was born in northern New Jersey, where doctors detected he had hypoglycemia (32-33 mg/dL) hours after birth. He was diagnosed with hyperinsulinism (HI) within 24 hours. When doctors there were unable to control his hypoglycemia with medication, he was transferred via ambulance to Children's Hospital of Philadelphia (CHOP) on day of life 8. He failed medical management, and results of a genetic test indicated diffuse HI due to homozygous mutation in *ABCC8*. R.A. underwent a near-total pancreatectomy at 17 days old. Hypoglycemia persisted, and he was discharged on octreotide during the day and dextrose via g-tube overnight.

He continued follow-up care at the Congenital Hyperinsulinism Center at CHOP. At age 2, octreotide was discontinued, in 2008 his dextrose was decreased, and in 2011 dextrose was discontinued. As is typical in patients who have had a near-total pancreatectomy, at age 12, he was diagnosed with diabetes with an A1C of 6.9%.

### Unique clinic created for unique patient population

R.A. was one of the first patients seen in CHOP's Multidisciplinary HI/Post-pancreatectomy Clinic, which was integrated further in 2017 to specifically treat this unique patient population in a way that was convenient for families — that no longer needed to make separate appointments with Endocrinology and Gastroenterology — and facilitated collaboration between clinicians to optimize coordinated care plans.

The clinic gathers an endocrinologist, gastroenterologist, dietitian and nurse practitioner who rotate through each patient's exam room. After all providers have assessed the patient, they huddle to develop or fine-tune the treatment plan, which is jointly reviewed with each patient and family. Referring physicians will receive one comprehensive follow-up letter that aggregates the input and care guidance from all specialists that have evaluated the patient during the clinic visit.

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In 2020, a genetic counselor was added to the clinic, as parents and older patients often have questions about the likelihood of future children having HI.

A neurodevelopment specialist will also be added to the clinic in 2020.

The clinic represents a collaboration between the HI Center in the Division of Endocrinology and Diabetes and the Pancreatic Disorders Program in the Division of Gastroenterology, Hepatology and Nutrition. The Pancreatic Disorders Program treats children with a wide variety of congenital or acquired conditions that impact the pancreas, from more common acute and chronic pancreatitis to much rarer presentations, regardless of cause. CHOP's HI Center has treated more than 1,300 children with congenital HI and performed more than 525 pancreatectomies — more than any center in the world. Its world-wide reputation has drawn patients from 48 states and 18 foreign countries.

## Missing pancreas, missing enzymes

One long-term complication of near-total pancreatectomy is exocrine/digestive pancreatic insufficiency (EPI). It's not a matter of if, but of when the child will require pancreatic enzyme replacement therapy. To track patients' potential need for pancreatic enzyme replacement, before coming to clinic, all patients have a single combined blood draw, which is tested for:

- A1C
- CBC
- CMP
- Lipid panel
- Fat soluble vitamins, vitamin B12 and essential fatty acid status

Patients also have a fecal elastase screening and have a DEXA scan to monitor bone mineral density. Without normal exocrine/digestive pancreatic function, children are at risk for multiple micronutrieint deficiencies that can be subtle but very consequential to a child's lifelong health and well-being. Our goal is to identify and screen all patients at risk, treat them and prevent any and all associated long-term potential consequences.

Families and referring physicians are prompted to contact CHOP if in between clinic visits the child shows symptoms of exocrine pancreatic inefficiency, which include: suboptimal growth, diarrhea; foul-smelling, greasy stools (steatorrhea); gas and bloating; stomach pain; hair loss; bleeding risk; and other signs, symptoms, and consequences of micronutrient and nutritional deficiencies.

## Tracking effect of octreotide

The Multidisciplinary HI/Post-pancreatectomy Clinic team also regularly follows and monitors HI patients with diffuse disease who are medically managed, as they may also be at risk for exocrine/digestive pancreatic insufficiency.

Generally, all children in the clinic are screened for poor growth, poor weight gain, and diabetes (if they have had a pancreatectomy). The dietitian works with families to ensure children are eating a healthy, well-balanced diet. Children with diabetes are counseled on avoiding sugary foods.

CHOP clinicians have learned that post-pancreatectomy patients of all ages can suffer from EPI. A handful of babies have been started on pancreatic enzyme replacement therapy before discharge post-surgery, some need it as toddlers, while others only require it closer to adolescence or later.

## Active, healthy, well-managed diabetes

At age 12, R.A. was started on pancreatic enzyme replacement therapy, which he takes before meals and snacks.

Now 15, R.A. manages his diabetes with a continuous glucose monitor and insulin pump, which are helping him keep his A1c in the target range. He is an honor roll student and is physically active, playing basketball and tennis. ■

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