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CASE REPORT CRSLS

Correction of Congenital Hyperinsulinism by Robotic-Assisted Laparoscopy in an Infant

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ABSTRACT

Background: Congenital hyperinsulinism (CHI) is a heterogeneous genetic disease characterized by increased insulin secretion, in which dysregulation of insulin secretion by pancreatic β cells causes persistent hypoglycemia in neonates and infants. Babies diagnosed with CHI require preferentially minimal invasive surgical treatment with near-total pancreatectomy (NTP).

Material and Methods: CHI was treated with robotic-assisted laparoscopy (RAL).

Results: The authors present an unreported case of CHI in an infant less than 10 kg, which was submitted to NTP treated by RAL. The procedure was performed with 3 arms of Da Vinci robot using adaptable size of trocars and the surgery was well succeeded.

Conclusions: The RAL is a challenge in pediatric pancreatic surgery to CHI due to the size of the trocars. To our knowledge, this is the first case reported in the English literature of an infant weighing less than 10 kg having been submitted to NTP by RAL.

Key Words: Complications, Congenital hyperinsulinism, Diagnosis, Follow-up, Robotics, Surgery.

INTRODUCTION

Congenital hyperinsulinism (CHI) is a genetic disease that demands immediate attention. It causes persistent hypoglycemia in neonates and infants, posing a significant health risk in this vulnerable population.[1] The incidence of CHI is 1 in every 40,000–50,000 live births in the general population, and paternal consanguinity can reach 1 in 2,500 live births.[2] CHI may have focal or diffuse or histopathological involvement of pancreatic β-cells and is predominantly caused by inactivating mutations in the two genes encoding the β-cell ATP-dependent potassium channel (KATP).[3] CHI is a neonatal emergency, as early diagnosis and management of hypoglycemia is extremely important to prevent permanent brain injury such as cerebral palsy, epilepsy, and death.[4,5]

Open surgery is the traditional approach to pancreatic resection when clinical treatment (diazoxide, glucagon, sandostatin) is ineffective. However, recent advances in laparoscopic surgery have led to laparoscopic near-total pancreatectomy (NTP) for diffuse lesions and laparoscopic distal pancreatectomy for focal lesions distal to the head of the pancreas.[6] We recently demonstrated the effectiveness of the minimally invasive surgery technique where laparoscopic pancreatectomy for medically unresponsive CHI is feasible and safe.[7] We report the first case of robotic-assisted surgery of an infant of CHI.

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Conflict of interests: none.

Funding sources: none.

Disclosure: none.

Acknowledgments: L.S. thanks the Sao Paulo State Research Foundation (FAPESP) for support Grant #2022/12021-1. The authors thank Ms. Carly Schafer for her assistance with English correction.

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DOI:10.4293/CRSLS.2025.00024

CASE REPORT

A seven-month-old, 8-kg infant girl, born and raised in Ribeirão Preto, Sao Paulo, Brazil. *Maternal data*: 45 years old, G2P1A0, underwent cesarean section due to fetal bradycardia. *Neonatal data*: Newborn was born at 38 weeks, Apgar 8 and 9, evolving in the maternity ward with hypotonia, swallowing disorder, and episodes of convulsive crisis that were controlled with the use of phenobarbital 5 mg/kg per day. She was hospitalized in another institution for 30 days after birth to investigate the convulsive condition, with a genetic syndrome being ruled out and being discharged for neurological follow-up. *Clinical investigation*: At 5 months of age, medical attention was sought after she started to have a runny nose associated with lethargy, and hypoglycemia (30 mg/dL) was then identified. During this hospitalization, glucose replacement was performed, and an investigation into an infectious disease was started, which was ruled out in the laboratory. The infant developed hypoglycemia, even after dietary adjustments, prompting the start of intravenous glucose infusion, which was quickly increased in order to maintain blood glucose levels above 70 mg/dL, up to a maximum dose of 26 mg/kg per minute. The critical sample showed blood glucose of 26 mg/dL, insulin of 16.7 IU/mL, ketonemia of 0.8 mMol/L, and negative free fatty acids. Once the diagnosis of hyperinsulinism was made, a sample was taken to perform an exome, though no changes were found, and there was no response to diazoxide at a dose of 15 mg/kg per day. Subsequently, a continuous SC infusion of Sandostatin was used up to 20 μg/kg per day, also without response. Blood glucose levels were better controlled with a continuous IV infusion of Glucagon without, however, achieving maintained blood glucose levels above 70 mg/dL in the absence of glucose infusion. *Surgical procedure*: The patient was classified as ASA II. The total time of anesthesia was 356 minutes, and the total time of surgery was 245 minutes. The child was positioned in the supine position with the abdomen elevated, and the robot was positioned to dock on the left side (**Figure 1**). The robotic-assisted NTP was performed using the Da Vinci XI robot, with the use of three robotic arms (arms 2, 3, and 4) on 8 mm/16 cm straight line trocars at the level of the umbilical scar, as well as with two auxiliary trocars, one of which was 8 mm in the right iliac fossa (RIF), and the other 5 mm in left iliac fossa (LIF) (**Figure 2**). Due to the size of the child, it was not possible to implant another trocar to use auxiliary arm 1 of the robot. A pressure of 12 mmHg with a flow of 18 mmHg was used to create a pneumoperitoneum. Pancreatectomy was performed according to the Kimura technique, with

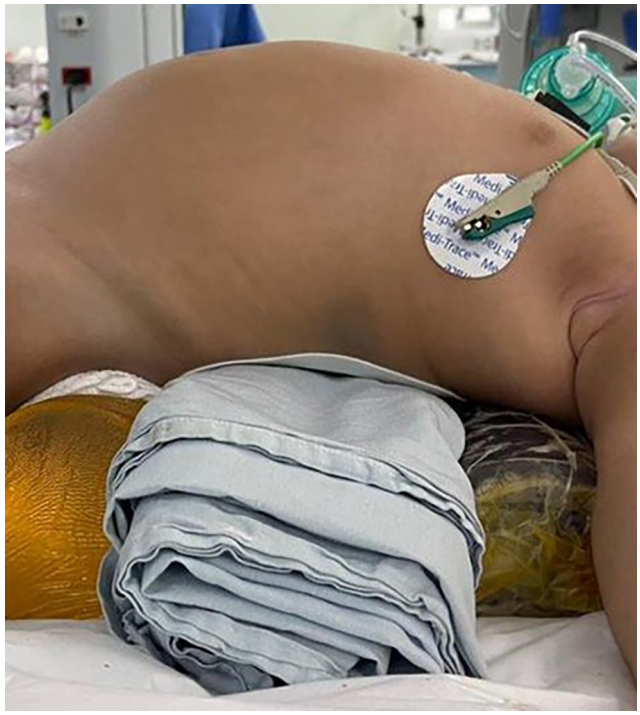

Figure 1. Positioning the patient for docking.

preservation of the splenic vessels and spleen, with the uncinate process of the pancreas being maintained after resection with a 30 mm, 8 mm diameter, curved-tip video laparoscopic stapler (**Figure 3**, overview). A Blake drain was maintained in the topography of the pancreas with a path in the left parietocolic gutter, exteriorizing at the site of the 5 mm trocar in the LIF.

Subsequent to NTP, an endoscopic gastrostomy was performed for alternative feeding due to a history of swallowing disorder and hypotonia. The patient remained stable throughout the procedure, free from hypoglycemia. The glucose saline was suspended intraoperatively. *Postoperatory and evolution*: Referred to the pediatric intensive care unit (PICU) for postoperative care. The patient's blood glucose level remained unchanged, requiring no corrections. Diet started on the fifth postoperative period after resolution of the adynamic ileus, with amylase collected from the drain fluid on the first, third, and fifth postoperative period remaining unchanged; however, on the fifth postoperative period, already in the infirmary bed, she presented with evisceration through the umbilical wound, requiring wall resuturing in the Surgical Center She developed acute respiratory stress syndrome (ARDS) progressing to pneumonia related to mechanical ventilation during the seventh postoperative period. She was discharged from the PICU on the 45th postoperative day vfor infirmary care. *Pathological anatomy result*: Biopsy

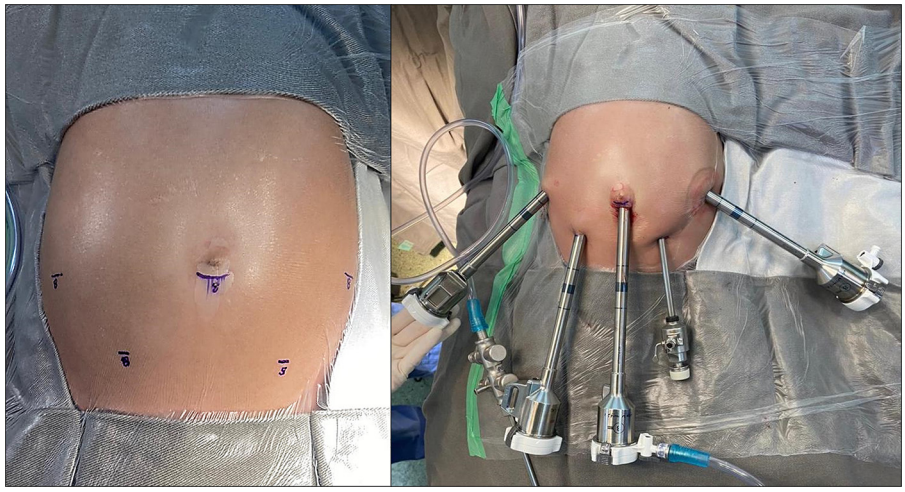

Figure 2. Positioning before and after the insertion of the trocars.

resulting from pancreatectomy identified diffuse CHI with microcystic ductal dilation and foci of proliferation at the small pancreatic ducts. No genetic mutations were found in the tissue.

DISCUSSION

The use of robotics in pediatric surgery has been evolving rapidly, with robots now being used for abdominal surgeries such as urological procedures,[8] anterior bowel defects,[9] choledochal cysts,[10,11] and splenectomies[12] in children. This field has shown rapid growth over the past few years, indicating a promising future.[13] Importantly, we are now observing a clear trend in which robotic surgery is increasingly being applied to pediatric surgical patients, approaching the same utility as in adult patients.[14]

The robotic platform requires pediatric surgeons to adapt robotic systems to children's surgical needs.[15] Furthermore,

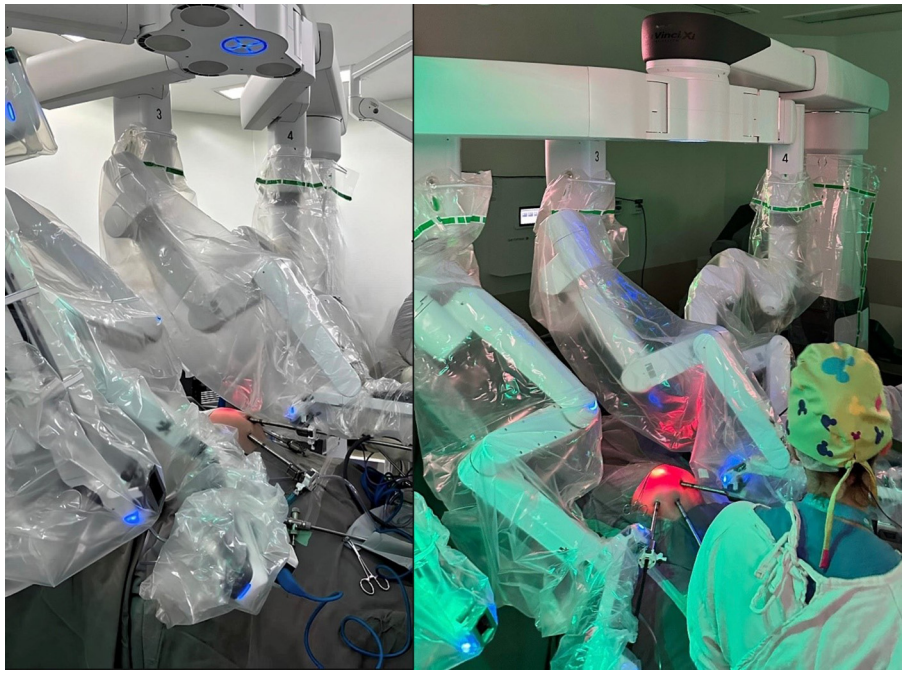

Figure 3. Overview of docking and portals.

smaller instruments and surgical techniques are needed to optimize the relatively limited workspace in babies and younger children, in addition to adapting the robot's nuances to the child's physiology.[16]

Since it was first performed in 2001, robotic pancreatic surgery in adults has become increasingly popular. Benefits seen with other types of robotic surgery, such as lower estimated blood loss and shorter length of hospital stay (LOS), are again seen in robotic pancreatic surgery. Some studies have started to associate robot-assisted distal pancreatectomy (RDP) and robotic pancreatoduodenectomy (RPD) with equivalent oncologic outcomes as compared to open procedures and even improved lymph node harvest.[17] Robotic total pancreatectomy (RTP) is a novel surgical approach currently performed by a select group of skilled surgeons. As robotic approaches to pancreatic surgery increase worldwide, rates of RTP are expected to increase.[18]

A case of near-total robotic pancreatectomy (NTP) for the treatment of nesidioblastosis after Roux-en-Y gastric bypass was initially performed in 2016 as an alternative to the treatment of complications of bariatric surgery.[19] The experience of utilizing NTP robotic pancreatectomy for the treatment of nesidioblastosis in children has not been described.

Tertiary centers have hundreds of CHI cases focused on multidisciplinary treatment,[20] and surgical treatment of pancreatic tumors is restricted to a few centers worldwide, with a high rate of over 15 patients per center.[21] The surgical treatment of CHI, comparing eight patients undergoing open pancreatectomy versus ten patients undergoing laparoscopically, showed a safe and significantly faster feeding time for the minimally invasive route, showing its safety and efficacy for most pediatric pancreatic tumors.[22] Our tertiary care center experience for CHI further supports these results that performing total or near to total pancreatectomy is effective in the treatment of tumors, with a resolution rate of 100% of patients.[7]

In terms of molecular genetics, 11 different genes are associated with various forms of CHI, most linked to monoallelic recessive KATP.[5–23] Only 9% of diffuse ICH may not have congenital causes, as in our patient.[20] Despite having robotic-assisted laparoscopy (RAL) for CHI resection, our patient also needed gastrostomy due to swallowing disorder and sequelae of multiple episodes of hypoglycemia that led the infant to neuro psychomotor development disorder (NPDD). These complications are not uncommon, as up to 75% of patients with CHI require feeding through a nasogastric tube or gastrostomy,[4] and 54% may have NPDD.[24] The early complication on the fifth post operative related to evisceration through the umbilical wound was related to the possible size of the trocar and, therefore, a complication related to the attempt to minimize the use of an additional robot arm.

In conclusion, CHI is a rare tumor that requires treatment by a multidisciplinary team. Once the diagnosis is confirmed, the indicated surgical treatment is preferably laparoscopic. While RAL has been increasingly used in adults, this is the first instance of a patient of less than 10 kg with CHI submitted to NTP performed by RAL. Advances in the miniaturization of robots will help expand the application of RAL in neonatal surgery.

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