

A simultaneous liver-pancreas transplantation in three years old patient with WRS

An anaesthetic case report from Karolinska University Hospital

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INTRODUCTION

Wolcott-Rallison Syndrome (WRS) is a rare syndrome affecting pancreas, liver, kidney and bone. It is an autosomal recessive disorder, with poor prognosis. Main clinical features are infantile onset insulin-dependent diabetes mellitus, multiple epiphyseal dysplasia, osteopenia, mental retardation or development delay, renal failure and recurrent acute liver failure following viral infections. Liver failure is a characteristic feature of WRS and most children die before they turn 10 years old. En bloc transplantation of liver, pancreas and kidney is technically a challenging procedure; however is the most effective option to increase survival as reported in the literature.

We present the anesthetic perspectives of liver and pancreas transplantation in a three years old child with WRS, at Karolinska University Hospital/Huddinge.

METHODS

Medical history

- A baby girl was born at full term as a first child to healthy, non-blood related Eastern European parents after normal pregnancy. The birth weight was 2675 g and the length was 47 cm. Uneventful postnatal care.
- At 5 weeks of age she was admitted to the local hospital in southern Sweden with high fever and severe dehydration. The Initial diagnosis was Diabetes Mellitus Type 1. Additional genetic testing confirmed the diagnosis of WRS.
- At the age of 6 months insulin pump and port-a-cath were implanted. Her general health improved and growth was within normal range.
- At 3 years of age, following several multidisciplinary team meetings, it was decided that 'en bloc' liver-pancreas transplantation would offer a better survival option.
- In February 2016, at age of 3 year she was referred to our transplantation center. She was preoperatively assessed by the pediatric transplantation team and listed for the transplantation.
- One month later her condition deteriorated rapidly, she ended up with an acute liver and renal failure following a severe upper airway infection and was admitted to PICU. The patient was intubated and mechanically ventilated and was diagnosed with severe RS virus infection. She received a 10-day CRRT.
- She recovered from acute multi-organ failure and discharged. Five months later a donor was found, and she was transferred to Karolinska University Hospital for a liver-pancreas transplantation.

METHODS (Cont)

Anesthetics management

- After oral pre-medication with Clonidine 3 microgram/kg she was calm entering the operation with her parents. She weighed 14 kg and was 97 cm in length. Anesthesia was induced with Atropine 0,01 mg/kg, Fentanyl 3 mg/kg, propofol 2 mg/kg, Rocuronium 0.6mg/kg. She was intubated nasally without any complication. Sevoflurane (1.2 MAC) used as anesthesia maintenance and intermittent Fentanyl 0.1 mg/kg. The patient was catheterized with two lumens wide bore jugular venous catheter and bilateral radial artery catheters. For echocardiography pediatric transesophageal probe was used. Blood analysis were done frequently and TEG.
- At surgical incision, Remifentanyl infusion was added due to high Fentanyl requirements. Noradrenaline infusion was used as a vasopressor. The Insulin pump was switched off and replaced by short acting Insulin infusion. Ringer Acetate and Albumin 50mg/ml was used as perioperative fluid therapy in fluid warming giving sets. Fluid Management System (FMS) was used for blood and fresh plasma transfusion, cell saver was used as well.
- Surgical piggy-back technique used for hepatectomy, rapidly increasing serum lactate with moderate hemodynamic instability during anhepatic phase. Adrenalin was administered, 0,01 mg/ml before recirculation. Mild reperfusion syndrome followed and lasted 8 minutes. It was noticed direct decline in serum lactate in phase 3. Heparin infusion administered after artery anastomoses. Pancreas transplantation procedure began after a short hemostasis period and underwent without any hemodynamic events, declining of insulin and Noradrenaline dosage at last hours of operation. The abdominal wall closed partially because of relatively large liver. The patient was transferred to the ICU anaesthetized.
- In postoperative day two, the patient had to be taken back to the theatre due to bile duct leakage and secondary approaching of abdominal wall was done. Patient's vitals were stable and liver function was improving, normal renal function test and no insulin need. Patient was extubated postoperative day 3 in ICU and continued in recovery pattern. Abdominal wall closure was done postoperative day 6 without any event. The patient was discharged to transplantations ward day 8 postoperative.
- The patient had vomiting and didn't accept nasogastric tube postoperatively, so nutrition was administered intravenously for a few days in the ICU and on the ward. However, jejunostomy tube performed on patient.
- Patient discharged 20 days post-operative in good and stable condition to pediatric ward in the local hospital for rehabilitation.

RESULT

- Our three years old patient underwent a liver-pancreas transplantation with no major complications perioperatively and early period postoperatively. The patient tolerated well general anesthesia, minimal hemodynamic instability during anhepatic phase with less than 10 minutes postreperfusion syndrome. Pancreas transplantation procedure went uneventfully. Total perioperative blood losses was 550 ml. The new organs function noticed promptly without any sign of early graft failure. Patient extubated postoperative day 3 in the ICU uneventful and delayed closure of abdominal wall postoperative day 6 and discharge to the transplantation ward day 8.
- The postoperative period was complicated by vomiting and bad tolerance of the nasogastric feeding tube, gastritis and hypertension. The patient received adequate treatment and jejunostomy performed. Immunosuppression's maintenance was provided with tacrolimus, mycophenolate mofetil and prednisolone. The patient discharge from our center to local hospital pediatric ward day 20 with a good stable health condition.
- Three years post transplantation, the patient is recovered well, enjoying normal daily activities, attending preschool. Liver and pancreas function are completely normal, remains Insulin free. However, renal function deteriorated with low GFR.

DISCUSSION

We report the first case of a simultaneous liver-pancreas transplantation for the management of life-threatening complications of WRS performed at Karolinska University Hospital without serious complications. In literature there are three cases reported before.

This procedure might offer better survival option and might prevent serious complications of WRS. One important success factor was our multidisciplinary team including pediatrician, pediatric hepatologist, endocrinologist, transplant anesthesiologist and intensivist, surgeons, specialized nursing staff.

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