TOTAL PANCREATECTOMY WITH ISLET AUTOTRANSPLANTATION (TP-IAT) IN CHILDREN - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Chronic pancreatitis and recurrent acute pancreatitis in children can lead to debilitating recurrent episodes of severe abdominal pain, significantly impact quality of life with dependence on opioid analgesia and can result in irreversible parenchymal injury to the pancreas. Repeated parenchymal damage can result in a slow progression to exocrine and endocrine insufficiency and eventually diabetes mellitus as islet cell mass is destroyed.
- Most children with chronic pancreatitis have a hereditary cause due to gene mutations or have anomalies of their pancreatic duct. In both these conditions, endoscopic or surgical treatments may not work. In addition, some genetic mutations are associated with a long-term risk of malignancy.
- Total pancreatectomy with islet auto transplantation (TP-IAT) is considered in select children with pancreatitis who have not or cannot respond to medical, endoscopic and /or surgical therapies and in whom the debilitating pain is an indication for a total pancreatectomy. Islets can be extracted from the removed pancreas and auto transplanted back into the child to significantly reduce the risk of complete insulin deficiency and diabetes post total pancreatectomy.
- This practice guideline provides guidance for diagnosis and selection of patients for total pancreatectomy and islet auto-transplantation for chronic pancreatitis.
- Furthermore, it provides medical and nursing staff at WSHLD and CHW with the medical and nursing management of the patient before, during and after the TP-IAT procedure.

Approved by:	SCHN Policy, procedure and Guideline Committee	
Date Effective:	1st May 2023	**Review Period**: 3 years
Team Leader:	Staff Specialist	**Area/Dept**: PICU - CHW

CHANGE SUMMARY

- Not applicable - new document
- **02/04/24** – Minor Update -section 7.3 – insulin infusion made up in normal saline, Appendix 2 BGL change in last hour the ranges of 0-5% changed to 2%. Format 7.1 to categories and lists. 7.1b updated to include more detail on MMTT. Appendix 5 Care plan template added.

READ ACKNOWLEDGEMENT

- Read Acknowledge Only – Nursing staff PICU and operating theatres, Department of Anaesthesia, Department of General Surgery Medical Staff, PICU Medical staff.

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1 Purpose

This document has the purpose to provide medical and nursing staff at and the Children's Hospital at Westmead (CHW) with the medical and nursing management of the patient who has undergone the TP-IAT procedure.

Total Pancreatectomy with Islet Auto-transplantation (TP-IAT) in chronic pancreatitis is a surgical procedure utilised in children with painful and debilitating chronic pancreatitis who have significant impairment to quality of life due to the pain, are at a risk of pancreatic insufficiency, diabetes, and pancreatic malignancy. Chronic Pancreatitis can result in a slow progression to exocrine and endocrine insufficiency and diabetes as islet cell mass is destroyed by repeated parenchymal injury. Islet Auto-transplantation following total pancreatectomy can significantly reduce the risk of severe hard to control diabetes post total pancreatectomy.

2 Intended Audience

The Children's Hospital at Westmead staff involved in the pre-operative management and post-operative care of the TP-IAT recipient.

3 Expected Outcomes

Management of Total Pancreatectomy and Islet Auto-transplantation for Chronic Pancreatitis within The Children's Hospital at Westmead.

4 Definitions

CP	Chronic Pancreatitis
RAP	Recurrent Acute Pancreatitis
IAT	Islet Auto Transplant
IBMR	Interstitial Blood Mediated Response
TP-IAT	Total Pancreatectomy with islet auto transplantation
TP	Total Pancreatectomy
HGP	Hereditary /genetic pancreatitis
IEQ	Islet cell equivalent
PRSS1	Protease trypsin 1
SPINK 1	Serine protease inhibitor Kazal type 1
CFTR	Cystic Fibrosis transmembrane conductance regulator
CTRC	Chymotrypsin C
ERCP	Endoscopic Retrograde Cannulation of Pancreatic Duct
PERT	Pancreatic Enzyme Replacement Therapy

5 Required Information

Pancreatic anatomy and function

The pancreas has two main functions: exocrine and endocrine. The greater portion of the pancreas is the exocrine function that is responsible for the digestion of carbohydrates, proteins and fats for the essential conversion to energy. The endocrine part consists of the Islets of Langerhans which produce insulin and the counter-regulatory hormone glucagon.

The normal human pancreas is a soft vascularised organ contained in a fibrous capsule. The approximate size of the normal pancreas in adult females is 84.88 +/- 14.95g and adult males 90.3 +/- 5.0g. There are approximately 1-1.5 million islet cells in the healthy human pancreas.

Pancreatitis in children

Acute pancreatitis can occur secondary to several causes such as trauma, stones, drugs, infections, immune causes, anomalies of the ducts and genetic causes. Most episodes of pancreatitis will settle although severe necrotizing pancreatitis can be serious.

Recurrent acute pancreatitis (RAC) is defined as more than one episode of acute pancreatitis. RAC may vary in severity and duration for each episode.

Chronic Pancreatitis (CP) is defined as the irreversible damage to the pancreas as a consequence of inflammation of the organ. Cellular, morphological and functional changes lead to pancreatic calcification with associated nutritional malabsorption, diabetes mellitus (pancreatogenic -type 3c) and pain syndromes.

The definition of CP in children was established by the INSPPIRE consortium in 2012 and requires one of the following:

a) Abdominal pain of pancreatic origin and imaging findings suggestive of chronic pancreatic damage.

b) Evidence of exocrine pancreatic insufficiency and imaging findings suggestive of chronic pancreatic damage.

c) Evidence of endocrine pancreatic insufficiency and imaging findings suggestive of chronic pancreatic damage.

d) Pancreatic biopsy specimen demonstrating histopathologic features compatible with chronic pancreatitis.

Aetiology and natural course of chronic pancreatitis in children

Chronic pancreatitis (CP) in children is a rare disease most often due to hereditary causes. In an INSPPIRE study, the majority (73%) of children with CP had identifiable genetic mutations - 50% of these are *PRSS1* gene with others being *CFTR*, *SPINK1*, *CTRC* (chymotrypsin C gene), *CPA1*, *TRPV6*. 33% of children had CP due to pancreatic duct obstruction – mostly pancreas divisum, 16% were toxic/metabolic and 13% autoimmune.

Children with CP usually present with recurrent pain which can be debilitating and significantly impact quality of life with recurrent hospitalizations, school absences, impaired interactions with family and peers, malnutrition and feeding challenges. The disease is usually progressive and increasing pain leads to narcotic dependence. Progression to

exocrine and endocrine insufficiency is common, including insulin dependent diabetes. Patients with CP due to hereditary/genetic causes have a high lifetime risk of pancreatic cancer.

6 Diagnosis and Selection of Patients

When is total pancreatectomy with auto-islet transplantation indicated?

Management of CP is aimed at reducing the episodes of pain, limiting dependency on opioid analgesics, and restoring quality and normalcy of life for the child.

While medical and endoscopic treatments are considered the initial treatment approach, total pancreatectomy (TP) is considered for debilitating CP in children who fail maximal medical and endoscopic management. Islet cell auto-transplantation preserves endogenous insulin release from beta cells. Thus, TP-IAT can provide durable pain relief in children, restoring quality of life and enabling them to achieve good glycaemic control.

The primary purpose of the procedure is to relieve pain and give the child a good quality of life and in addition, mitigate long term risks of pancreatic cancer. The secondary outcome is to salvage all available islets with the hope of retaining native islet function. Often, if there is impaired glucose tolerance or existing diabetes, this may be harder to achieve.

Criteria for considering TP-IAT in children

Two large volume units use the following criteria for considering TP-IAT

Cincinnati Children's Hospital criteria for TP-IAT in children

a. Pain or debilitation for at least 6 months, with either chronic
    i. opioid dependence or impaired quality of life due to frequent
    ii. school absence and/or hospitalizations.
b. Failure of maximal medical and endoscopic therapies.
c. Absence of reversible cause of CP or ARP.
d. Absence of medical or psychosocial contraindication.
e. Willingness to accept risk of lifelong diabetes.
f. Non-diabetic status, IFG, IGT and/or C-peptide positive diabetes

Minnesota Criteria for TP-IAT in children

1. Abdominal pain of > 6 months duration with impaired quality of life e.g., inability to attend school, inability to participate in ordinary activities, repeated hospitalizations, or constant need for narcotics, each coupled with failure to respond to maximal medical treatment or endoscopic pancreatic duct drainage procedures.
2. In addition, there must be objective findings of CP, including at least one of the following:
    a) pancreas calcifications on CT scan, or abnormal ERCP, or ≥ 6/9 criteria on endoscopic ultrasound (EUS);
    b) or (2) any two of following:
        (1) ductal or parenchymal abnormalities on secretin stimulated magnetic resonance cholangiopancreatography (MRCP), EUS of pancreas with 6/9 criteria positive, or abnormal pancreatic function tests with peak bicarbonate < 80 mmol/L).;
        (2) Histopathologic confirmed diagnosis of chronic pancreatitis from previous operations;

(3) Hereditary pancreatitis (PRSS1 gene mutation, SPINK1 gene mutation, CFTR gene mutations, CTRC or otherwise), with a compatible clinical history;
(4) History of recurrent acute pancreatitis with > 3 episodes of pain associated with imaging diagnostic of acute pancreatitis and/or elevated serum amylase or lipase 3 times normal[17]

Timing of TP-IAT in hereditary pancreatitis

Patients with CP due to hereditary/genetic causes are unlikely to respond to medical or endoscopic treatments and have a high lifetime risk of pancreatic cancer. TP-IAT should be considered early in the course of the disease when there is less pancreatic fibrosis, more viable islets and a higher chance of greater islet yield. Moreover, several studies have demonstrated significant improvement in quality of life and decreased dependence on opioids in patients with hereditary/genetic CP.

Impact of previous procedures on outcome of TP-IAT

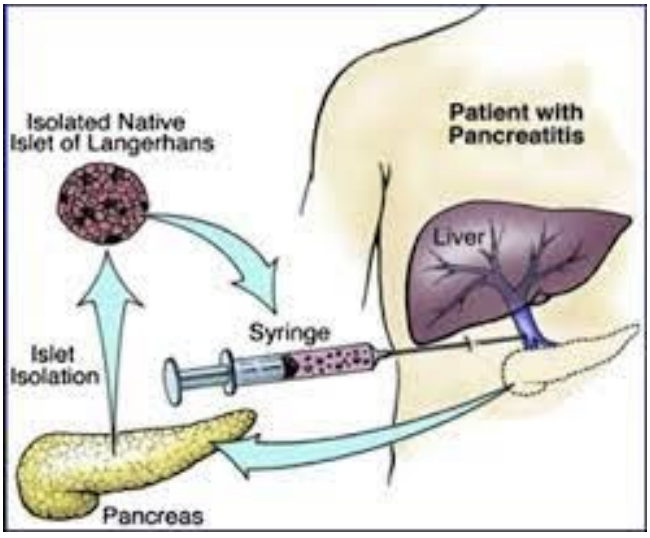
Previous surgical drainage procedures (Puestow or Frey's) compromise islet yield if a subsequent TP-IAT is done. In view of this, one paradigm is to do any indicated drainage procedures primarily by endoscopic methods alone with limited use of traditional surgical drainage, keeping in mind endoscopic procedures may compromise islet yield.

Impact of age on outcomes of TP-IAT

Children less than 11 years of age have better outcomes with a greater proportion becoming insulin independent post islet auto-transplantation.

7 Overview of the procedure

Total pancreatectomy is performed with techniques to minimize warm ischemia time in order to preserve islets. Once the pancreas is removed, it is immediately flushed with preservative solution to maximize islet preservation. The pancreas is then transported to the islet processing facility at Westmead where they are isolated and prepared for auto transplantation into the patient. Once processed, islets are then re-infused into the patient's own liver where they settle and grow. As it's the patient's own cells, immunosuppression is not required.



7.1 Pre-Surgical Assessment

Confirmation that Pancreatitis is the primary diagnosis. Review by surgical, gastroenterology and endocrinology teams at CHW and islet team (including surgical) at Westmead hospital. This is the checklist of all the actions needed in the preoperative assessment.

1. **Blood tests**
    a. Genetic panel – look for hereditary pancreatitis: panel should include *SPINLK1*, *PRSS*, *CTRC*, *CFTR*. Optional genes include *CPA1* or additional genes listed onPanel App.
        - https://panelapp.agha.umccr.org/panels/154/
        - If the patient has a genetic cause for pancreatitis found, cascade testing of the family members should follow and a pedigree documented.
    b. Assessment of endocrine / islet reserve –
        - Mixed meal tolerance test (MMTT) using Boost High Protein HP, 6mL/kg, to maximum of 360mL, with glucose, insulin, and C-peptide levels at 0, 30, 60, 90 and 120 minutes. If Boost HP is not available dietetics can advise a suitable substitute.
        - HbA1C
        - Insulin autoantibodies - specifically IAA, GAD, IA2 and Zn-8 auto antibodies
    c. Routine bloods – FBC, EUC, LFTs, Coagulation profile

2. **Medical imaging**
    a. CT scan
    b. MRI scan
    c. Ultrasound – assess Portal venous patency

3. **Immunization for potential asplenia** –
functional or anatomical (i.e., splenectomy) refer to SCHN Immunisation practice guideline.
Patients with an absent or dysfunctional spleen are at life-long increased risk of fulminant bacterial infection, most notably invasive pneumococcal disease.
Recommended vaccinations for all patients with asplenia are27:
    a. Pneumococcal
    b. Meningococcal
    c. Hib (Hiberex)
    d. Influenza

    Resource:
    Australian Government Department of Health and Ageing – Table - Recommendations for vaccination in people with functional or anatomical asplenia: https://immunisationhandbook.health.gov.au/resources/tables/table-recommendations-for-vaccination-in-people-with-functional-or-anatomical-asplenia

For patients undergoing elective splenectomy, vaccination should be completed, where possible, 2 weeks prior to the scheduled operation date. If unplanned splenectomy occurs, vaccination should occur approximately 1 week after surgery.

4. Consultations

a. **Paediatric endocrine consultation**,
- Including evaluation of family history for diabetes and autoimmune disorders.
- Evaluate pre-morbid glycaemia by determining presence of impaired fasting glucose (IFG), impaired glucose tolerance (IGT) or diabetes and the premorbid insulin secretion response to mixed-meal tolerance test (MMTT) with insulin/C-peptide levels and HbA1c.
- Measurement of auto antibodies which may predispose to the development of type 1 (immune mediated) diabetes should also be performed pre-operatively: specifically IAA, GAD, IA2 and Zn-8 auto antibodies

b. **Diabetes educator**
- Post-operative insulin administration via multiple daily injections (MDI) or pump with continuous glucose monitoring (CGM) may be required, and pre-operative education should be commenced.
- Insulin pump and CGM device should be ordered and available prior to the procedure

c. Dietician
- to include carbohydrate counting for dosing of insulin and fat counting for subsequent dosing of Creon.

d. Pain Team
- Assessment preop with plan for post operative pain management: Early management of pain with optimal therapy targeting the aetiology rather than symptoms. Referral to the Pain Clinic at The Children's Hospital at Westmead for pre-evaluation of pain management and post-operative pain management.

e. Psych Med
- If required

f. Anaesthesia
- Preoperative assessment via pre-admission clinic. The anaesthesia department is notified early and is part of planning MDTs.

g. Paediatric Intensive Care
- The team is notified early in the planning process, is part of MDTs and the family have the option of meeting them prior to surgery.

h. Islet transplant unit at Westmead adult hospital
- The team is notified early in the planning process, is part of MDTs and the family have the option of meeting them prior to surgery.

i. Gastroenterology team
- A gastroenterologist is often the primary referring physician, they are part of the MDTS and actively involved in the management of these patients pre and post op

5. Consent signed for Surgical Total Pancreatectomy and Auto Islet Cell Transplantation with or without splenectomy.

Pathology /Investigations: Blood samples taken at initial consultation and pre-operatively.

Vacutainer Required	Test
Light purple (EDTA)	FBC
Light purple (EDTA)	HbA1c
Light Green	EUC CMP Urate Iron Profile LFT Amylase Lipase Lipid (Cholesterol, Triglyceride, HDL,LDL) Blood Glucose, Bicarbonate
Gold Top	C-peptide, insulin IAA, GAD, IA-2 and ZnT8 (if not done previously)
Pink (with Black Centre)	Cross Match
MSU	Ward urinalysis to observe for: Blood Protein Glucose Ketones Leukocytes Nitrates

7.2 Pre-Operative

Additional important consultations:

- Anaesthesia – See Appendix 1 for anaesthesia workup and intraoperative protocol.
- PICU: See Appendix 2 for
  - PICU protocol
  - Individualized/ customized template for ICU management.
- Pain team assessment if indicated.
- Child life therapist if indicated.
- Hematology – for 10 bottles of 20% albumin

7.3 Medication Orders

NO PREDNISOLONE IS USED FOR ISLET TRANSPLANTATION AT ANY TIME – AS STEROIDS ARE ASSOCIATED WITH ISLET DEATH AND LOSS

Operative (induction) Medication Orders

Medication	Dosage
Pipercillin + Tazobactam	IVI: 100 mg/kg (Max 4 g) stat
Narcotics	As per patient preoperative dose
Heparin	70 units/kg divided in 2 doses as follows i. 35 units/kg added to islet cell preparation ii. 35 units/kg administered at time of guide-wire insertion
Insulin	Starting insulin rate is 0.025 units/kg/hr IVI (to maximum starting rate 2 units per hour) Insulin infusion to be made up in normal saline not dextrose Insulin infusion-to commence at time of ligation of pancreatic vessels discontinuing endogenous insulin delivery

7.4 Surgical Management

7.4.1 Surgical Equipment Setup Requirements

- 10 x 100 mL Human Albumin 20%, 20g in 100mL CSL Biotherapies
- Operating table to allow for radiological imaging at time of line insertion
- Harmonic Scalpel or Ligasure
- Thomson/ Omnitrack Retractor
- Sodium Heparin total 70 units/kg
- Infusion Catheter TERUMO GLIDECATH COBRA 2 (C2) FR5 GM maximum guidewire to use with it 0.38”(0.97mm)- confirm availability with Dr Wayne Hawthorne
- High Pressure Connecting tube
- Pressure monitoring Transducer
- Double male adaptor x2
- Standard Bore Extension Set
- Contrast Media
- Glucometer with Glucose and Ketones testing strips
- Endo GIA stapler and the bipolar for the intestine and haemostasis

7.5 Anaesthesia considerations

Discussion about heparinisation and risks. This should happen prior to start of anaesthesia – discuss need for intraoperative anticoagulation with heparin. Discussion should include review of TEG, coagulation status, safety of epidural and postop pain management.

- **NO STEROIDS to be given at any time.**
  - This is very important as **steroid use is associated with islet death and must not be used.** (Many anaesthetists would routinely use dexamethasone as an antiemetic in this scenario and this should be avoided).
- Analgesia and pain management
  - Epidural: this is the preferable method of management with these children given chronic pain issues and high risk of severe acute pain postoperatively
  - **Although a large dose of heparin will be given as a bolus intraoperatively, this is not expected to happen till at least 4 hours into the procedure so placing the epidural should be safe.**
  - Alternative strategies would be intrathecal morphine or with wound catheters plus PCA oxycodone/ other agents
- Blood sugar monitoring, timing and insulin management
  - Expect a sudden surge in insulin release when the pancreas is being handled and dissected with a risk of hypoglycaemia at that time.
  - Once the pancreas is out, BSLs are more labile and it is important to ensure strict euglycemia especially while the islets are being infused and thereafter. It's very important to have very tight sugar control to avoid stressing the islets which can lead to islet loss.
  - Prepare and connect an insulin infusion once central line is in with hourly measurements if stable to start with.
  - **Once islets are ready** to be infused, do 5 minutely BSLs just before and during the infusion of the islet cells. This is then switched to 15-minute measurements.
  - See protocol attached in Appendix 2 and 3
  - Ketones as per your protocol. Logistically this means getting some ketone sticks.
- Other areas of concern / interest
  - NGT and CVL at the start
  - Avoid nitrous due to bowel distention
  - One arterial line should be fine but some anaesthetists may prefer 2 due to need for frequent blood sampling.

7.6 Surgical Procedure

Total pancreatectomy

In paediatric patients this should performed via a midline or a bilateral subcostal surgical incision for the pancreatectomy. Larger children and adult patients may benefit from laparoscopic mobilisation of the pancreas, thereby reducing the size of the incision. The pancreas is mobilized preserving its blood supply until the dissection is completed for resection, thus minimizing the warm ischemia time and maximizing islet cell preservation. Papaverine may be used to avoid spasm to the arterial supply to the pancreas.

If possible, the pylorus is preserved to minimize biliary reflux. Preservation of the spleen is preferable in children. It should be preserved if preserving it doesn't compromise warm ischemia to the pancreas.

Just prior to clamping vessels to remove the pancreas, **30-50 units/kg of heparin can be given IV to aid in preserving blood flow to the pancreas.** Some units give this to improve perfusion to pancreas just prior to vascular clamping in order to better preserve the pancreas and improve islet yield. (Note: **This is in addition to** the 70 units/kg of heparin that will be given into the portal vein approximately 4 hours later when the islets are infused). Heparin has a half-life of 60 -90 minutes and it is anticipated that very little would be left in circulation in 4 hours' time when islet infusion begins.

Although removing the pancreas whole results in better islet yield, if necessary, the neck of the pancreas can be divided, and the pancreatic head is removed separate from the body and tail to minimise any ischaemic injury to the islets prior to processing.

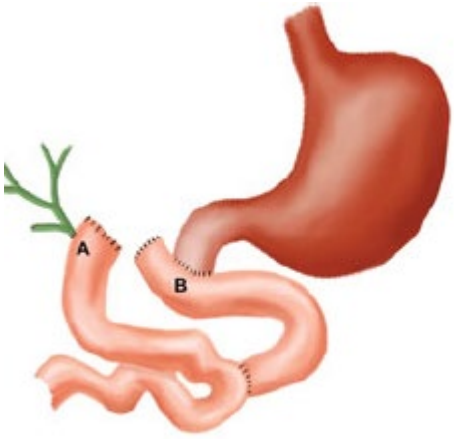
Islet isolation

The pancreas is removed and transferred to the Islet Processing Team where the Islet cell isolation procedure will be performed. This is anticipated to take 2-4 hours. The islets are isolated by collagenase digestion of the pancreas and mechanical digestion using the method described by Camillo Ricordi. Islets are usually separated from excessive acinar tissue by density gradient centrifugation. However, in children, this is hard to achieve, and the islet cells are not fully purified by density separation and are not cultured following the isolation process. Some acinar tissue remains with the islets, but this doesn't seem to matter too much in children. The islets are suspended in albumin and prepared for infusion and auto-transplantation. A culture is taken from the suspension.

Gastrointestinal reconstruction

While the islets are being isolated, reconstruction of the patient's gastrointestinal tract and bile duct is performed. Although there are many methods of reconstruction, with a pylorus preserving approach, the jejunum can be anastomosed to the duodenum and a 40 cm roux loop used for biliary drainage.

Most children have post-operative gastroparesis and establishing feeds and nutrition is often a problem. A gastrojejunal feeding tube can be placed via a gastrostomy in order to establish continuous jejunal feeding via the J tube while venting the stomach via the G tube.



The patient will remain anaesthetised and ventilated while the isolation procedure is being performed with the abdomen packed while waiting for islet isolation to be completed.

Blood glucose monitoring during pancreatectomy and immediately after completion of pancreatectomy: See Anaesthetic intraoperative management – See 9.2.1

During manipulation of the pancreas and just prior to its removal, blood sugars can fall precipitously due to an insulin surge released by distressed islets. Following removal of the entire pancreas, complete absence of endogenous insulin can result in significant hyperglycemia.

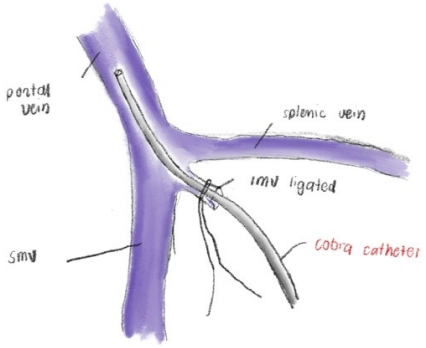
Bloods sugars monitoring during this time needs to be frequent (initially every 5 minutes) with appropriate measures to achieve euglycemia.

Once the pancreas is removed, BGLs should be monitored every 15 minutes via finger prick or via arterial line. If BGL > 15 mmol/L, blood ketones should also be measured. Refer to anaesthesia protocol – appendix 2 for detail protocol.

Insulin glucose infusion should commence at time of ligation of pancreatic vessels when endogenous insulin delivery ceases.

Preparation for Islet infusion

The inferior mesenteric vein is isolated, and the infusion catheter placed into the portal vein and secured as shown. A 3 way cannula is attached to one end in order to measure portal pressures intermittently while the islets are being infused.



7.7 Islet infusion

Calculating volumes for infusion.

Portal vein infusion of auto transplanted islet cells can be complicated by portal vein thrombosis, bleeding and transaminitis due to the non-purified nature of the transplanted cells. Tissue volume (TV) / kg body weight of islets predicts risk of complications and therefore a TV/kg of less than 0.25 mL is considered a safe volume to infuse.

A maximum packed cell volume (tissue volume) of 0.25 mL per Kg body weight will be infused. The standard suspension volume for paediatric cases is 120 mL per bag with 100 mL rinse with a 220 mL suspension volume. For small children or where there is a concern regarding the fluid volume, the suspension volume is reduced to 100 mL (w/ 50 mL rinse)[26].

Maximum volume of islet prep to be infused:

Maximum amount of islet prep the aim at putting in less than 10 mL of digested pancreatic tissue in total. Preps exceeding this volume will require the COBE isolation step to reduce the volume

Total volume of transplant media to be infused with the Islets:

Standard protocol of 120 mL of transplant media is used as the carrier agent for the Islets then a further 50-100 mL of transplant media is used as a flush/wash to ensure all islets are washed out of the transplant bag into the patient

IBMIR (Immediate blood mediated inflammatory reaction) and risk of portal vein thrombosis

Intravascular infusion of islets triggers a severe, non-specific inflammatory response—immediate blood mediated inflammatory reaction (IBMIR)—which is responsible for much of the islet loss that occurs (around 50 % of the islets can be lost during this time). In addition, there is a risk of portal vein thrombosis.

To reduce the risk of portal vein thrombosis and to mitigate the IBMIR response, a total of 70 units/kg of Heparin is given - 35 units/kg administered directly into the portal vein just prior to islet infusion and 35 units/kg added to the Islet Cell Preparation[11].

Heparin dosing

Total of 70 units/kg (dose is calculated using recipient weight)

Divide doses:

i. 35 units/kg patient body weight in bag to prevent aggregation of the tissue

ii. 35 units/kg patient body weight administered immediately prior to time of Cobra wire insertion into Portal vein

Portal Vein Pressure measurements

Portal venous pressure is measured at 5 minutely intervals whilst the islets are infused. Reduce infusion rate if Portal pressure increases from the previous reading by more than 5mmHg[12].

If the Portal Pressure monitoring reaches 20 mmHg (25 cm H_20), or a total tissue volume of greater than 10 mL of tissue is infused (packed cell volume), the infusion is discontinued and remaining infusion placed in the peritoneal cavity[1].

7.8 Completion of Surgical procedure

Following infusion, a thorough check for haemostasis is made, along with a check for splenic viability. Drains are usually placed and the abdominal wound closed. 15 Fr Blake drains work best. If there is no epidural, wound catheters can be placed for post op pain relief.

8 Post-Operative Care

Patient should go straight to Paediatric ICU escorted by the anaesthetist. Extubation may occur in theatre prior to transfer to PICU. Patients go directly to PICU post operatively, not via recovery.

8.1 Key Considerations in Post-Operative Management

- **Tight Glucose control**

  Tight blood glucose level (BGL) control is vital to ensure the newly transplanted islet cells survive as hyperglycaemia and swings in BGLs can result in islet cell loss. Management of blood glucose levels will follow the protocol as outlined and will be principally supervised by the endocrinologist on call in conjunction with the intensivist.

- **Anticoagulation**

  Post-operative heparinisation is given to ensure portal vein thrombosis does not occur and to reduce inflammation. Heparin is given as continuous IV infusion at 10 units/kg/HOUR for at least one week post operatively.

- **Pain Relief**

  Most children with CP have chronic pain and resultant anxiety. Managing pain and anxiety could be challenging.

- **Nutrition**

  Post-operative gastroparesis is a very common problem and most high-volume centres place a gastrojejunal tube for continuous feeding post operatively, TPN will be started day 2 post operatively and continuous jejunal tube feeds should be commenced by day 4 or when gut motility is established. This strategy makes it easier to manage both the nutrition and obtain the required tight blood sugar control in PICU.

Nursing care of the patient is 1:1 nurse patient ratio.

- The nursing staff should notify the Registrar/CMO when informed by the theatre staff of the patient's impending return.

- The anaesthetist will accompany the patient on their return from theatre. Check the following with the anaesthetist:
  - The operative course
  - Analgesia administered.
  - Reversal agent and time/effect of administration
  - Total estimated blood loss in theatre
  - Fluids given in theatre.
  - Current blood glucose level and rate of insulin and glucose infusion
  - Orders for further analgesia/ Pain Team Management
  - A nasogastric tube is used for gastric decompression to prevent nausea and vomiting.

8.2 Nursing responsibilities

Should observations fall "outside the flags" appropriate PACE & ALS protocols are to be adhered to[15].

- **Blood Glucose Monitoring to occur as per TP-IAP protocol. 15 minutely for 2 hours then every 30 minutes for 4 hours, then hourly while on IV insulin**
- Insulin and glucose infusions to be titrated in response to BGL's as per protocol (appendix 2)
- Patient to be placed on continuous cardiac and pulse oximeter monitoring with hourly documentation of vital signs to include:
  - Blood Pressure
  - CVP (whilst indicated via distal CVL lumen)
  - Heart rate
  - Respiration rate
  - Pulse Oximetry values
  - Pain scores
- PCA care and observations – if in use.
- Wound Drainage monitoring, documentation and replacement of drain loses mL/mL
- Gastric aspirate monitoring and documentation
- Catheters (urinary) Management as per practice guideline
- Formal Bloods - patient should continue to have early morning (09:00hours) formal blood tests for tests listed in the table below the duration of their hospitalisation to aid in the planning of their care.

Vacutainer Required	Blood Test
Light purple (EDTA)	FBC
Light Green	EUC LFT
Grey	BSL
Blue	COAG
Gold Top	C-peptide Day 3 and day 7 post operatively while on IV insulin in PICU. Once transitioned to a subcutaneous insulin pump and still in hospital, measure stimulated C-Peptide once weekly (one hour post meal)

8.2.1 Patient Assessment and Diagnostics

"Should observations warrant, appropriate PACE & ALS protocols are to be adhered to[11]

- Points to check during assessment and review include:
  - **i.** Blood samples need to be taken for urgent EUC, LFT, FBC, BSL, and coagulation studies.
  - **ii.** Chest x-ray to check position of the central line and presence/absence of pneumothorax. X-ray to be reviewed by medical officer.
- The main surgical risk of the procedure is either liver haemorrhage or infarction of the liver. Therefore, if pain scores increase significantly and/or hypotension or tachycardia occurs, an urgent liver ultrasound and LFT are indicated with an urgent FBC and Hb.

8.2.2 Other management guidelines relevant to this procedure

- **i.** Catheters (urinary) Management Procedure - The Sydney Children's Hospitals Network Procedure
- **ii.** Intravenous Fluid Management –CHW Practice Guideline

Post-operative feeding and insulin requirements

Immediate post operatively, the patient is nil by mouth with a NG tube to free drainage that is aspirated and discarded 4 hourly. If the patient has a GJ tube, the gastrostomy is left on free drainage.

TPN is to be commenced on day 2 post operatively and IV insulin administration continues as per endocrinology.

Continuous jejunal tube feeding is commenced when gut motility is established and the surgical, endocrinology and ICU teams are comfortable with starting feeds. This is usually by Day 4. Feeds can be graded up and titrated with TPN. The gastric tube can be vented and left to free drainage if there is gastroparesis.

It's important to ensure tight sugar control during all this especially during the transition.

While on IV insulin, insulin adjustments are made according to the Paediatric ICU Algorithm for Insulin Adjustments for TP-IAT (appendix 2). Frequent testing of the blood glucose level occurs over the duration of care with insulin adjustments made hourly or at the discretion of the Endocrinologist or treating Physician.

Once the patient is tolerating an oral diet, the IV insulin infusions- transitioned to multiple daily subcutaneous injections or subcutaneous insulin pump therapy under the guidance of the Paediatric endocrinologist and /or diabetes educator /Nurse Practitioner.

Transition from IV Insulin to Subcutaneous insulin may alternatively depend on enteral tube feeding. Once the patient is stabilised on continuous enteral feeds at a stable and appropriate rate, transition to subcutaneous insulin can occur. This is typically around day 5-7 post-operatively[4]]. Oral diet may be commenced with decreasing amounts of gastric aspirate.

8.3 Peri and Post-operative insulin Infusion for islet transplant

Setting up and management of an insulin and glucose infusion- commenced at time of ligation of pancreatic vessels discontinuing endogenous insulin delivery.

- Syringe pump must be used for the IV insulin infusion.
- No other IV fluids/medications are to be added to insulin or glucose infusions. These two infusions should be run via a three-way tap on the blue lumen of the CVL.
- **Before attaching to the patient, prime the line with prescribed insulin infusion**. Leave for a minimum of 1 minute and then flush the solution from the tubing (4 mL) replacing it with fresh solution from the syringe. This allows the medication to coat the tubing and prevent absorption which may affect the dose of medication delivered to the patient. (See Appendix 3 – Management of Hypoglycaemia)
- If cannula tissues or occlusion occurs, BOTH the insulin and glucose infusions must cease, and alternate access gained as a priority
- Titration of as per Appendix 2: Perioperative and PICU IV Insulin Infusion Algorithm Post Total Pancreatectomy with Islet Cell Transplant.
- After the first 6 hours if there is ongoing recurrent hypoglycaemia requiring repeated cessation of insulin delivery increase the glucose concentration by 25-50% above the current infusion concentration to achieve adequate glucose levels to allow continued insulin delivery and prevent ketosis. Continue to titrate insulin as per Appendix 2.

8.3.1 Monitoring and observations for insulin infusion

- Patient will remain on insulin-glucose infusion for 5-7 days to maintain optimal glycaemic control for engraftment. An insulin pump or multiple daily injections usually commence when the patient is able to eat adequately to maintain euglycaemia or is on full enteral feeding protocol. This is to be a joint decision of the surgical and endocrinology teams. Subcutaneous insulin pump therapy should not be commenced until the patient is discharged from PICU to Clancy Ward. (Note: patient should only be transferred to Clancy ward).
- Islet function has been observed at 3 days, but most will not be of any benefit for glycaemia until >14 days post-transplant. Insulin requirements are often increased in the immediate post-operative period due to surgical stress and cytokine release.
- It is expected that total insulin requirements will decrease slowly with progressive engraftment and function of transplanted islets. Close monitoring is necessary to avoid very low BGL. **Frequency should be up scaled if islet lysis causes ongoing hypoglycaemia most likely in the first 4-6 hours post islet transplantation**.
- Note: **Ischaemic islets can release large amounts of insulin into the portal circulation, putting the patient at high risk of hypoglycaemia. Thus more frequent BGL monitoring is needed in the first 4-6 hours when this is most likely to occur**.
- **Intra-operatively:** BGL monitoring to be performed every 5 minutes following infusion of islets.

- **Post operatively:** BGL monitoring to be performed every 15 minutes for 2 hours, then every 30 minutes for 4 hours, then hourly until 48 hours post TP-IAT.
- Monitor blood ketones 4 hourly in first 24 hours post-transplant. Notify Endocrinologist if blood ketones more than 1 mmol/L. Ketones should also be tested if insulin infusion is ceased for 30 minutes for hypoglycaemia correction.
- **Insulin rate to be adjusted hourly based on BGL unless BGL is less than 4.5 mmol/L**.
- **Recommence insulin infusion as per Paediatric ICU TP-IAT protocol (Appendix 2) when BGL is more than 5 mmol/L**
- **Manage blood glucose as per the Paediatric ICU TP-IAT Protocol (Appendix 2)**

8.4 Pancreatic Resection Post-Operative Complications

- **Gastric Ileus –**Post prandial nausea and vomiting, decreased bowel sounds, constipation, anorexia.
- **Endocrine Insufficiency-** Hyperglycaemia and associated symptoms
- **Fistula-** Abdominal pain/distension, fever, chest pain, increase naso-gastric secretions.
- **Biliary Reflux-** Chronic acid reflux, bile emesis, dyspepsia, tooth decay
- **Exocrine Insufficiency-** Post prandial hyper peristalsis and diarrhoea, malnutrition secondary to lack of fat absorption, weight loss
- **Diabetes requiring insulin.**
- **Lifelong need for Pancreatic Enzyme Replacement** to prevent fat malabsorption in the duodenum.
- **Post Splenectomy** complications - Changes in blood and platelet counts rendering the patient more susceptible to infection. Immunisation recommendations (Vaccination Protocol for Paediatric patients with functional or anatomical asplenia)
- **Dumping syndrome**

8.5 Pain Management

- Assess and evaluate pain location and quality, document as part of routine care. Document pain severity and sedation scores hourly while epidural or opioid infusions (PCA/NCA) are running. Pain scores (at rest and on movement) together with 'opioid-usage' can usefully guide analgesic prescription[8].
- Minimal discomfort is associated with percutaneous trans-hepatic and laparoscopic transplant procedures. If pain is moderate to severe underlying causes should be explored.
- Pain Relief, Morphine or Fentanyl infusion as required, or oral therapy as tolerated as per the pain team, who should be consulted. Paracetamol can be used if the liver function is good (10mg/kg per dose Q 4- 6 hourly, with a maximum of 4 doses per 24 hours)[19].

8.6 Central Line Management

Losses of abdominal drains should be replaced millilitre for millilitre with attention to CVP and urine output. Aim for CVP 8mmHg depending on urine output. Lost fluid can be assessed for its electrolyte and albumin content so that appropriate fluids can be used for fluid replacement (usually normal saline and/or 4% albumin). Monitor blood values including serum sodium, urea, creatinine, haemoglobin and haematocrit to assess for intravascular depletion[19].

Patient will return with a triple lumen central line to be connected as follows[11, 20]

Distal lumen (Brown)	• Central venous pressure (CVP) if required
Medial lumen (Blue)	Insulin and glucose infusions via single three way tap (no chooks foot), no other injection port to avoid inadvertent bolus.
Proximal lumen (White)	• Medications and maintenance fluids • Heparin infusion
PIVC	• Analgesia (PCA) can be infused via a PIVC

Note: Fluid replacement should aim to keep patient euvolemic thus maintaining normal CVP levels of 8 mmHg

- **Risk of infection**. Central venous line may become infected typically days 5-7 post procedure. Fever at this time should prompt consideration of line removal and appropriate antibiotic therapy (usually Vancomycin)[19]

8.7 Other Tests

- Doppler Ultrasound of the liver[19]
  - Within 24 hours of returning to ward (High dependency or PICU)
  - If reverse portal venous flow is noted, the Surgical Team should immediately be notified.
- Chest x-ray on admission to PICU, after 12 hours then daily while the endotracheal tube is in place.
- Cultures:
  - From Day 2, then second daily until three lots of negative results
  - Bacterial cultures of sputum, urine, abdominal drains and bile.
  - Viral Cultures if indicated.
  - MRSA Screening swabs of nose, groin, rectum on PICU admission and then weekly and on discharge from PICU
  - Ensure that all drain tips are cultured on removal.

8.8 Medications

- Aspirin (soluble) is given once daily when feeds are established, as prophylaxis against Portal Vein thrombosis. Dose 5mg/kg/day up to maximum 50mg (half 100mg tablet)

Patient is to remain NBM until surgical and endocrine review[19].

Medication	Indication	Dosage
NBM:		
Insulin	Hyperglycaemia	Initial starting dose is 0.025units/kg/hour via intravenous infusion up to a maximum starting rate of 2 units per hour, then review rate hourly according to Insulin adjustment algorithm in appendix 2
Piperacillin + Tazobactam	Prophylaxis	100mg/kg/dose Q8 H IV
Heparin	Thromboprophylaxis	initial: 10units/kg/hr via continuous intravenous infusion given for 7 days.
When feeding:		
Aspirin	Thromboprophylaxis	Aspirin (soluble) is given once daily when feeds are established, as prophylaxis against Portal Vein thrombosis. Dose 5mg/kg/day PO up to maximum 50mg
Insulin	Hyperglycaemia	Continue IV insulin as basal insulin while in PICU. When introducing feeds liaise with Endocrine team for advice about meal bolus insulin
Amoxycillin	Asplenia Prophylaxis	20mg/kg/dose (max 250mg) PO daily for 12 months in patients with splenectomy
Pain Medication	Pain Management	Refer to Pain team
Omeprazole	Ulcer prophylaxis	1mg/kg/dose PO once daily
Fat Soluble Vitamins A, B12,D,E,K	Supplement	Vitamin ABDECK: 4-10years: 1 capsule daily PO >10years 1-2 capsule daily PO Vitamin A, D and E: 0-12 months: 0.7mL PO daily 1-3 years: 1mL PO daily

- **PANCREATIC ENZYME REPLACEMENT THERAPY Dosing Recommendation for the treatment of Paediatric Doses[22] (See Appendix 4)**

	Starting Dose	Titration Considerations	Minimum to Maximum Dose
Meal	1000 to 2000 lipase units/kg/meal	Adjust dose to control steatorrhoea and maintenance of nutritional status	500-2500 lipase units/kg/meal
Snack	Half of meal dose		Half meal dose

1 scoop Creon Micro (5000units lipase) per 3/6 g fat (WSLHD – Dietetics 2013)
1 Creon 10 000 (10 000units lipase) per 6-8 g fat
1 Creon 25 000 (25 000units lipase) per 15-20 g fat
1 Creon 40 000 (40 000units lipase) per 25-32 g fat
The dosage may vary in the development of patient gastrointestinal symptoms such as diarrhoea, and weight loss. The doses are titrated upward to alleviate the symptoms[13] .

8.9 Follow Up Management and Education

- Avoid heavy lifting for 6-8 weeks post operatively[1.]
- Insulin therapy – Following discharge, insulin doses are to be adjusted by the CHW endocrinologist in conjunction with the CHW diabetes nurse practitioner and senior diabetes educators. If the patient comes from another Local Health District, AND has access to a tertiary trained paediatric endocrinologist, the CHW endocrinologist may consider transferring care back to the local endocrinologist / diabetes team or a shared care model can be negotiated on a case-by-case basis,
- Insulin should be adjusted in accordance with blood glucose levels and weaned if possible. Suggested review periods are weekly for 1 month, then 2-4 weekly until 6 months, then 2 monthly until 12 months post-transplant.
- Duration of insulin therapy is anticipated to be for at least 3 months duration. After 3 months slowly wean meal coverage and then basal insulin as tolerated. (Bellin, 2018). Note: If the patient is on a Hybrid Closed loop pump the auto basal will adapt to the reduced insulin requirements with time from transplant, but the carbohydrate ratios will need to be progressively weakened by the diabetes team as insulin requirements decrease.
    - Amoxicillin antibiotic therapy for 12 months post-splenectomy
    - Follow up 3, 6 and 12months and thereafter annually, post-surgery to include:
        i. Pathology investigations: HbA1c
        ii. Measurement of stimulated C-peptide at each visit (non-fasting, 1 hour post meal)
        iii. Serum measurement of liver chemistry (LFT, GGT, ALT)
        iv. Nutrition management and management of Exocrine Insufficiency Assessment of steatorrhea, weight monitoring, fat soluble vitamins levels.
        v. Adjustment and education to patient and family/ carer of exocrine medication, Education diabetes management
        vi. Pain team management
        vii. For patients who undergo splenectomy the paediatric vaccination protocol needs to be instituted (Vaccination Protocol for Paediatric patients with functional or anatomical asplenia)

8.10 Ward Guidance

Post operative medical and surgical management of the child will follow the same protocols/ guidelines as in the document.

Blood sugar control, insulin requirements, feeds, surgical management will be closely directed by the endocrinology team, surgical team and the gastroenterology teams

For all questions and matters related to glycaemic (blood sugar) control – please call the endocrinologist on call. For all other matters, the surgical team.

9 Education Notes

- The whole pancreas is resected. The patient's own pancreatic Islet cells are isolated from the pancreas and infused into the liver via the portal vein. The cells are grafted onto the liver, which is then responsible for the secretion of insulin in these patients.
- No immunosuppression is required for these patients as they are receiving their own Islet Cells.
- Insulin therapy is required following the procedure and is weaned under the management of the Endocrinologist.
- Approximately 40% of patients undergoing the procedure achieve insulin independence.
- A large percentage of patients experience pain relief and show a significant improvement in Quality of Life[23]

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Appendix 1: Heparinisation Protocol for Paediatric Auto Islet Transplantation

Preoperative

- Thromboelastography (TEG) done in theatre during induction
- Coagulation profile PT/PTT and INR available from preoperative labs.

Intraoperative

If deemed suitable at the time of the islet transplant 35 units/kg of Heparin given IV via the intra portal catheter at the time of placement. If the Islet/acinar suspension is deemed to be more procoagulant then the heparin will be all added to the islet bag prior to infusion. Additional 35units/kg heparin will be added to the bag with islets which will already contain 35units/kg of heparin making a total of 70 units/kg of heparin that the patient will receive.

- TEG can be done 1 hour post infusion.
- Check anti-Xa and start Heparin continuous intravenous infusion at 10 units/kg/hr 2-hours after islet cell infusion if TEG is OK, and no significant bleeding.

Post-operative: In the PICU

- Continue heparin infusion and usually aim for antiXa 0.2 – 0.4.
- Organise Doppler US of liver at 12-24hrs post-op to confirm NO EVIDENCE OF PORTAL VEIN CLOT
- Continue Heparin infusion FOR 7 DAYS with daily Anti Xa and TEG
- Target Anti Xa between 0.2 and 0.4 usually considered sufficient but should be confirmed daily following discussion with surgeon.
- Aspirin 5 mg/kg PO (up to a maximum of 50mg) given once daily can be started on day 3 once epidural catheter is out and feeds commenced and tolerated.

Appendix 2: Algorithm for Insulin Adjustments (TP-IAT)

Not to be used in Diabetic Ketoacidosis or Hyperglycaemia Hyperosmolar Syndrome. Only to be used in paediatric ICU.

- Calculate change in infusion rates based on the hourly blood glucose level as per APPENDIX 2 [see below] (BGL, even in situations where blood glucose is being measured more frequently immediately post-surgery)
- If BGL less than 3.3 mmol/L, **follow red box instructions on the TP-IAT protocol.**
- If BGL greater than or equal to 3.3 mmol/L, follow **algorithm adjustments.**

Insulin is not commenced pre-operatively unless the patient has previously received exogenous insulin therapy. Insulin commences at time of clamping of pancreatic blood vessels.

Appendix 2 (continued): Perioperative and PICU IV Insulin Infusion Algorithm - Post Total Pancreatectomy with Islet Cell Transplant

Starting insulin rate is 0.025 units/kg/hour continuous intravenous infusion (to maximum starting rate 2 units per hour) using 1 unit/mL solution in PICU. i.e. 50units/50mL 0.9% NaCl.

Commence 5% glucose at maintenance rates when BGL <15 mmol/L.

The table indicates the **percentage change in insulin infusion rates** from the current hourly rate according to the current BGL and the rate of change of BGL in the previous hour. Note post islet transplant BGL may be tested more frequently than hourly but the calculation is to be based on the change in the last one hour excepting when hypoglycaemia occurs. The goal is to a maintain **target blood glucose of 5.5-6.7 mmol/L** which is critical for successful islet cell engraftment in the first 3-5 days post-transplant. If the blood glucose target range cannot be maintained, the endocrinologist on call should be contacted. Adapted from University of Minnesota Post op TP-IAT insulin infusion protocol.

<table>
<tr><th rowspan="2">Current BGL mmol/L</th><th colspan="5">Change in BGL from last hour</th></tr>
<tr><th>⬇ > 3</th><th>⬇ 1.6 - 3.0</th><th>⬇ 0.1 - 1.5</th><th>⬆ 0 - 1.6</th><th>⬆ > 1.6</th></tr>
<tr><td>>8.8</td><td>⬇ 10%</td><td>No change</td><td>⬆ 25%</td><td>⬆ 25%</td><td>⬆ 50%</td></tr>
<tr><td>7.3-8.8</td><td>⬇ 10%</td><td>⬇ 5 %</td><td>⬆ 25%</td><td>⬆ 25%</td><td>⬆ 25%</td></tr>
<tr><td>6.8-7.2</td><td>⬇ 10%</td><td>⬇ 10%</td><td>⬆ 10%</td><td>⬆ 10%</td><td>⬆ 10%</td></tr>
<tr><td>5.5-6.7</td><td>⬇ 10%</td><td>⬇ 10%</td><td>⬇ 2%</td><td>⬆ 2%</td><td>⬆ 10%</td></tr>
<tr><td>5-5.4</td><td>⬇ 25%</td><td>⬇ 25%</td><td>⬇ 5%</td><td>No change</td><td>No change</td></tr>
<tr><td>4.5-4.9</td><td colspan="2">Cease 30 mins, recheck BGL >5 then restart 75% previous rate</td><td>⬇ 25%</td><td>No change</td><td>No change</td></tr>
<tr><td>3.3-4.4</td><td colspan="5">Cease insulin until BGL >5 then restart 75% of previous rate</td></tr>
<tr><td><3.3</td><td colspan="5">Cease insulin. Give IV glucose 2mL/kg of 10% Dextrose.
Recheck BGL every 15 mins until BGL >5. Restart insulin at 50% of previous rate</td></tr>
</table>

Appendix 3: Management of Hypoglycaemia

Current BGL < 2 mmol/L or severe symptoms/signs of hypoglycaemia	Current BGL 2.1 to 3.3 mmol/L or severe symptoms/signs of hypoglycaemia
⇨ Cease insulin ⇨ Bolus 2mL/kg of 10% glucose and repeat at 15 mins if no response ⇨ Call **ALS or MET** if unconscious Repeat BGL measurement every 15 minutes ⇨ Resume insulin at 50% of previous rate when BGL ≥ 5 mmol/L ⇨ Do not increase insulin rate again for at least one hour after its commencement ⇨ Change to glucose 10% infusion if needed to reduce volume of glucose	⇨ Cease insulin ⇨ Bolus 2mL/kg of 10% glucose and repeat at 15 mins if no response. ⇨ Repeat BGL measurement every 15 minutes. ⇨ Resume insulin at 50% of previous rate when BGL ≥ 5 mmol/L ⇨ Do not increase insulin rate again for at least one hour after its recommencement.

- Recommence insulin infusion as per algorithm only when BGL is above 5 mmol/L

Appendix 4: Pancreatic Enzyme Replacement Therapy (PERT)

Are there any foods and fluids that don't need PERT?

Enzymes need to be taken with ALL food and fluids. The exception is foods that contain mainly simple sugars. Some examples include:

- Fresh, dried and canned fruit
- Non - starchy salad vegetables e.g. celery sticks,
- Lollies, jellies, sorbet, Roll-ups
- Fruit juice, cordial, soft drink, electrolyte replacement drinks, water

Remember that while enzymes are dosed according to how much fat is in food, enzymes work on fat, carbohydrate and protein. Therefore, they are still needed with foods that are lower in fat such as rice cakes, lean meat, baked beans, plain spaghetti and rice and egg whites.

When should I take the enzymes?

Take pancreatic enzymes immediately before meals/snacks. Enzymes last in the small intestine (where they work) for around 30 minutes. If you take longer than 30 minutes to eat, give half the dose at the start of the feed and the rest half-way through.

How should the enzymes be stored?

Store enzymes in the fridge or in a cool, dry place. Keep enzymes in an opaque container and out of direct light. Discard any enzymes that have been in the sun or left out in the heat e.g. in hot car.

Do not put enzyme beads in tube feeds, or down nasogastric tubes or gastrostomies.

Things I can do to improve managing my PERT:

1. _____

2. _____

3. _____

For further information contact your CF Dietitian – Sarah McKay 0428916296

Modified from: Consensus document from Dietitian/ Nutritionists from the Nutrition Education Materials Online, "NEMO", team. Disclaimer: http://www.health.qld.gov.au/masters/copyright.asp
Reviewed: December 2013 Due for review: December 2015

Fat Counter

Item	Household Measure	Weight (g)	Energy (kJ)	Fat (g)
Meat/Meat Equivalents				
Steak, panfried with oil	1 medium	120	1320	18
Roast Beef	2 slices	80	540	5
Lamb Chops (loin) panfried with oil	2 untrimmed	150	1670	26
Roast Lamb	2 slices	90	710	6.5
Roast Chicken with skin	2 slices	60	550	8
Wiener schnitzel (crumbed & panfried)	1 medium	180	1630	25
Bacon				
Grilled	1 rasher	50 (raw)	405	7
Fried	1 rasher	50 (raw)	470	9
Sausages				
BBQ	2 thin	100	1130	20
Salami	5 slices	40	480	10
Luncheon meat	1 slice	30	250	6
Fish, crumbed, fried	1 average	100	940	11
Egg				
Boiled/Poached	1	60	375	7
Fried	1	60	575	12
Takeaway & Fast Foods				
Hamburger				
Big Mac	1	250	2115	25
Cheese Burger	1		1220	12
McDonalds French Fries	Medium		1700	22
Chicken Nuggetts	6	114	1275	18
Chicken, crumbed & fried	1 average	130	1800	28
Meat Pie	1 average	175	1880	24
Sausage Roll	1 large	130	1570	28
Dim Sim - Fried	1	70	670	7
Pizza Supreme	2 slices	200	2400	26
Fish & Chips - deep fried				
Fish battered	1 piece	150	1590	23
Chips	20 medium	100	880	15
Fats				
Margarine or Butter	1 tspn	5	150	4
Oil	1 TBS	20	735	20
Cream	1 TBS	20	280	7
Salad Dressing	1 TBS	20	210	3
Mayonnaise	1 TBS	25	350	7
Peanut Butter	1 TBS	20	525	10

Item	Household Measure	Weight (g)	Energy (kJ)	Fat (g)
Milk & Milk Products				
Full cream milk	1 cup	250ml	675	10
Low fat milk (1.2-1.4%)	1 cup	250ml	550	3.5
Skim/non fat milk	1 cup	250ml	365	0
Milk powder				
Full cream	1 TBS	10	195	2.5
Skim/non fat	1 TBS	10	150	0
Icecream	1 scoop (100ml)	50	375	5
Yoghurt - flavoured	1 tub	200	790	5
Cheese	1 slice	20	350	7
Fruche	1 tub	200	960	8
Breads & Cereals				
Bread	1 slice	30	275	1
Dry biscuits e.g salada	1 biscuit	15	255	1.5
Plain sweet biscuits	2	30	660	7
Cream filled biscuit	2	30	660	7
Doughnut - Plain	1	70	1045	10
Croissant	1 medium	50	670	12
Cake - Plain	1 piece	60	940	11
Breakfast Cereal				
Cornflakes	1 cup	30	430	-
Weetbix	2	30	420	1
Muesli toasted	1/4 cup	30	500	3
Rice	1 cup	160	770	0.5
Fried rice	1 cup	165	1500	14
Spaghetti	1 cup	150	690	0.5
Spaghetti with bolognese sauce	2 cups		2425	14
Snack Foods				
Potato Crisps	Multipak	21	440	6.5
	Medium	50	1045	15
Popcorn	Medium bucket	55	1150	16
Peanuts	1/4 cup	30	1150	16
Muesli bar	1 average	30	520	4
Chocolate	6 squares	30	670	9
Mars Bar	1 regular	60	1165	11
	1 fun size	25	470	5
Soft Drink	1 can	375	675	-
Fruit/Vegetables				
Apple	1 medium	150	270	0
Banana	1 medium	150	365	0
Grapes	Small bunch	120	315	0
Orange	1 medium	280	335	0
Sultanas	2 T	30	385	0
Beans	1/2 cup	60	50	0
Broccoli	2 florets	45	45	0
Carrots	1 medium	140	135	0
Corn Cob	1 medium	100	200	0.5
Peas	1/4 cup	40	85	0
Potato				
Boiled	1 medium	90	300	0
Roasted	1 medium	90	473	5
Avocado	1/2 medium	80	535	13

Developed by WSAHS Repiratory Ambulatory Care Service, December 2002.
Acknowledgment to "Nutrition Information for Adolescents and Adults" The DAA Cystic Fibrosis Special Interest Group (Vic), 2001

Appendix 5: TP-IAT Individualised Patient Care template

TP-IAT: TRANSPLANT INDIVIDUALISED TREATMENT PLAN

PATIENT DETAILS: *NAME:* *MRN:* *DOB:*

DAY OF Tx= DAY 0

HEIGHT: | WEIGHT: | BSA: | CMV STATUS: | EBV STATUS: | BLOOD GROUP: | | CONSENT:

BLOOD PRODUCTS REQUIRED:

PERIOPERATIVE MEDICATIONS: **Pre-med: N/A**

Medication	Patient dose	Comments
Insulin Infusion		As per TP-IAT guideline section 8.3

POSTOPERATIVE MEDICATION

Medication	Patient dose	Comments
Insulin Infusion		As per TP-IAT guideline section 8.3

Microbiology prophylaxis tailored to patient:

Medication	Patient dose	Comments

Microbiology prophylaxis: Maintenance

On Call Endocrine

Day 0	Day 1	Day 2	Day 3

Special comments: