Short Bowel Syndrome and Intestinal Failure: Living Longer, Living Stronger

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Disclosures
• I have no conflicts of interest
• I will discuss the drug Gattex which is still in clinical trials by the Shire corporation.

How many of you work or have worked with patients with intestinal failure or short bowel syndrome?

What is Intestinal Failure?
The inability of the small bowel to ingest, digest and absorb sufficient nutrients, water and electrolytes in order to maintain the nutritional status of adults and provide growth and development in children.

Causes in Children
Too Little
• Gastroschisis
• NEC
• Intestinal Atresia
• Mid-gut volvulus

Does not function
• Pseudobstruction
• Microvillus Inclusion Disease
• Agangliosis
• Mitochondrial disorders
Intestinal Failure + Complications

- Central Line Infections
- Chronic diarrhea
- Gastritis
- Malnutrition and development
- Liver disease/GI bleeding
  - Skin telangiectasias
  - Feeding aversions
  - Porto hypertensions

Loss of central venous access
- Unable to provide IV nutrition or hydration
- Leads to Malnutrition, Electrolyte Imbalances
- Blood clots and altered perfusion

Transplant Indications

- Jan 2005 - 7mo with Gastroschisis referred for transplant
  - ~50cm intestine, ½ colon
- Advanced liver disease with splenomegaly
  - INR 1.5, platelets 33, bilirubin 12.5
- Both gastroenterologist and surgeon agree “will definitely need liver-small bowel transplant”
  - Listed as status 1 for Liver and Intestinal Transplant
- Continued Intestinal Rehab while waiting for transplant
  - Lipid reduction, advancement of feeds, central line care, optimized TPN, diet education
- May 2005 – dropped need for intestine transplant
- June 15, 2005 – made inactive for liver transplant
  - Continue aggressive rehabilitation in Nebraska
- Aug 28, 2009 – removed G-tube
- Eats and drinks 100% by mouth

Outcome

- Healthy 14 year young man in high school
- Clinic visit, DEXA scan and labs once a year
- Following his growth curves
- Remains off IV and enteral support
- Thriving academically
- Avid basketball/soccer player
- Mad gaming skills
- Big brother to 3 sisters

2005

Complete Rehabilitation
5 year total $402,129

Liver-Intestine Transplant
5 year total $1,444,370

5 Year Cost Savings
$1 Million

... work like this?

Can you make this...
Path for Intestinal Failure

Medical Management

- TPN reduction with enteral/oral advancement
- Early introduction to elemental enteral feedings
- Short gut diet
- Ethanol/Antibiotic Locks
- Vitamin/Mineral management
- Bone Age/DXA scans yearly
- Referral to a SBS/IF specialty center
- Teenage transition program
- Feeding therapy
- Intralipid minimization
- Bacterial/Fungal Overgrowth
- GLP-2 and other growth factors
- Teaching and Support
n=49 children
mean F/U 254 +/- 160d
PRE-ethanol
8.8 infect/1000days
POST-ethanol
3.4 infect/1000days
62% reduction (p<0.01)
If increase dwell time beyond 2h, rate to 2.1/1000days

Outcomes from EtOH Locks
Serial Transverse Enteroplasty

**STEP**

- Serial Transverse Enteroplasty

**Other Surgical Options**
- Fistula Repair
- Ostomy Takedown
- Formation of Ostomy
- Resection
- Tapering Enteroplasty

**Intestinal Transplant**

- **Indications**
  - Loss of venous access
  - End stage liver disease
  - Life threatening CVC infections
  - Non-reconstruct able GI tract

**1 WEEK**

**2-3 WEEKS**

**2-3 MONTHS**

**2-6 YEARS**
Stats
• Intestinal Transplant Survival Rates:
  • 75-85% at one year (SRTR data)
  • 60-70% at five year
• Lifetime immunosuppression to prevent organ rejection
• Increased risks for cancer and other rare illnesses
• Possible graft failure or loss leading to re-transplantation
• Despite this, many people are living full lives today following intestinal transplantation
• Goal is to take in nutrition and hydration 100% by mouth or feeding tube

Intestinal Transplants per Year

End of Transplant?
• Liver Disease
  • Omegaven/SMOF has improved liver condition/function
  • Risk of GI bleeding from varices minimized improved liver function
• Central Line Infections
  • Ethanol Lock solutions have minimized central line infections
  • Yeast and gram negative organisms falling – these were the most life-threatening
• Loss of venous access
  • Fewer line infections led to decrease in loss of central lines and need for replacement

Listed Patient Deaths

Pediatric Deaths Waiting for Transplant
No Limits

• Liver disease no longer scares us
• 50 cm? Why not 40…why not 30…20…10...or zero?
• Surgery is always an option…but just because we can, does not mean that we should!
• Give the gut a chance…no matter the length or function
• Don’t believe everything that you hear!
• Transplant is always the last resort, but is always an option

“Those who are crazy enough to think they can change the world usually do.”
- Steve Jobs

Questions?