

# Short Bowel Syndrome and Intestinal Failure:

## Living Longer, Living Stronger

Brandi Gerhardt, BAN, RN  
Intestinal Rehabilitation Program  
Transplant Coordinator  
Nebraska Medicine



# 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
- Pseudoobstruction
  - Microvillus Inclusion Disease
  - Aganglionsis
  - Mitochondrial disorders




## Intestinal Failure + Complications

- Central Line Infections**
  - Chronic diarrhea
  - Vomiting
  - Malabsorption
  - Poor growth and development
- Liver disease/GI bleeding**
  - Skin breakdown
  - Feeding aversions
  - Portal hypertension
- Loss of central venous access**
  - Unable to provide IV nutrition or hydration
  - Leads to Malnutrition, Electrolyte imbalances
  - Blood clots and altered perfusion


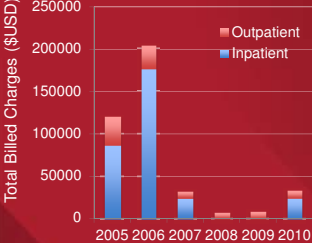
**= Transplant Indications**



# 2005




- Jan 2005 - 7mo with Gastroschisis referred for transplant
  - ~50cm intestine, 1/2 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





Year	Inpatient	Outpatient
2005	~80,000	~20,000
2006	~180,000	~20,000
2007	~30,000	~10,000
2008	~10,000	~5,000
2009	~10,000	~5,000
2010	~20,000	~10,000

Complete Rehabilitation  
5 year total \$402,129



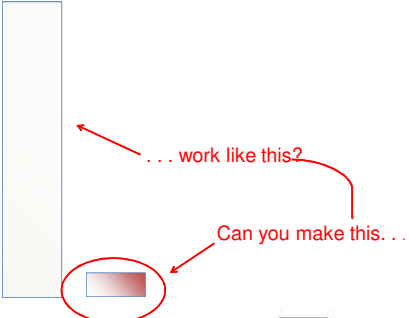

Liver-Intestine Transplant  
5 year total \$1,444,370

**5 Year Cost Savings  
\$1 Million**



## 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








# 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/DEXA 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





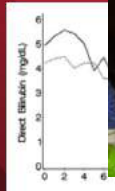
Parents hope fish oil can spare son's liver



“Omega baby w...  
“You sliced...  
July 2006  
... 2009  
07

Experimental supplement will replace soy liquid that could kill baby born with intestinal failure

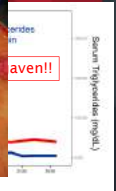





Diast. Bloodin (mg/dL)

Week


Omegaven



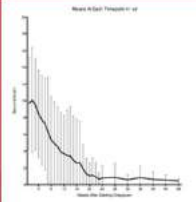
Serum Triglycerides (mg/dL)

Weeks

Lipid Minimization




# Omegaven Clinical Trial (n=53 kids)



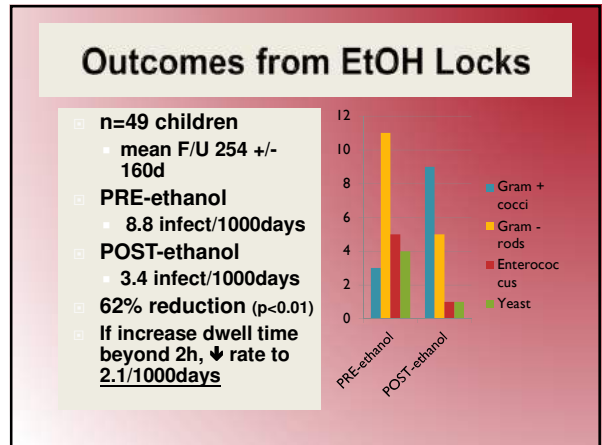
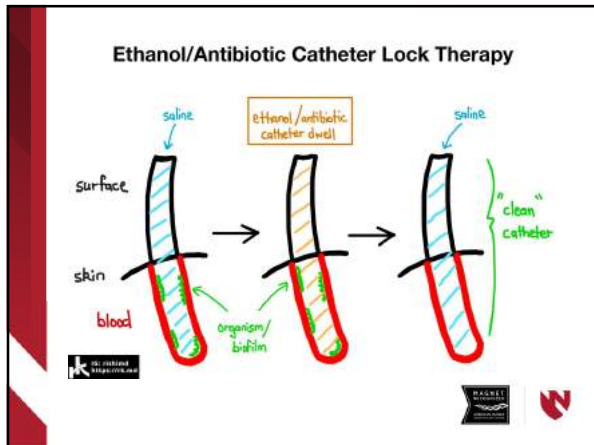
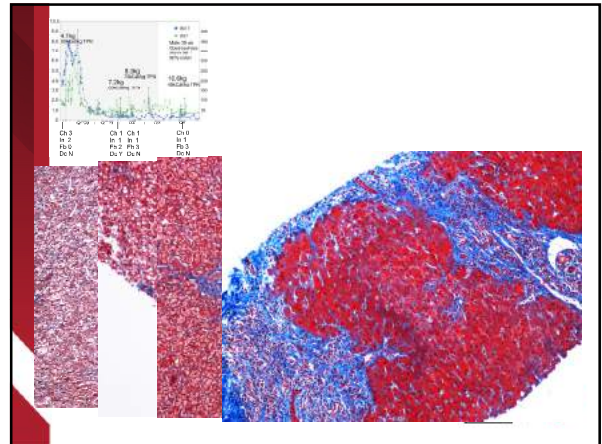
Omegaven

Soy Minimization



# Lipid Minimization



## Gattex

Enterodocrine Cell, Subepithelial Myofibroblast, Enteric Neuron of Submucosal or Myenteric Plexus

GLP-2 Receptor

Tecughitide

## Enteroplasties

~~Lengthening Procedures~~



### Bianchi Procedure

Bianchi, 1980 J Ped Surg 15(2):145-51

## STEP

- ▣ **Serial Transverse Enteroplasty**
- ▣ Described in 2003 by Kim *et al* (Boston Children's) in J Ped Surg 2003; 38: 425-9

### Other Surgical Options

- ▣ Fistula Repair
- ▣ Ostomy Takedown
- ▣ Formation of Ostomy
- ▣ Resection
- ▣ Tapering Enteroplasty



COMPLETELY <b>1 WEEK</b>
INPATIENT REHABILITATION Surgical correction of the underlying condition <b>2-3 WEEKS</b>
TRANSITION TO OUTPATIENT INTENSE OUTPATIENT REHABILITATION <b>2-3 MONTHS</b>
ONGOING REHABILITATION AT HOME Lifelong comprehensive nutritional management <b>2-6 YEARS</b>
COMPLETE ENTERAL TOLERANCE

## Intestinal Transplant

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

## 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



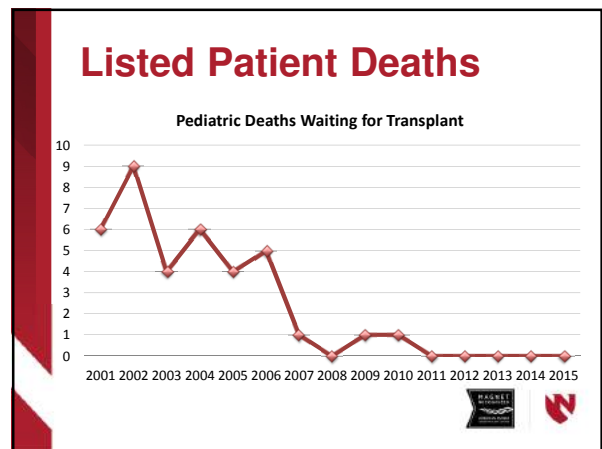
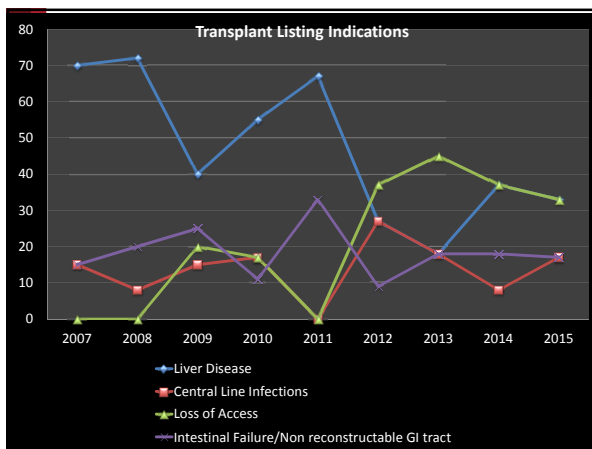
	2014	2013	2012	2011	2010	2009	2008	2007	2006	2005	2004	2003	2002	2001	2000
< 1 Year	3	1	9	8	7	22	29	37	31	26	23	10	15	20	10
1-5 Years	17	25	33	22	37	56	49	56	50	45	53	45	30	35	30
6-10 Years	6	5	8	2	10	11	10	8	6	15	10	7	10	5	7
11-17 Years	2	5	6	3	8	5	5	10	9	10	6	11	13	3	3
18-34 Years	9	12	18	13	25	21	34	27	34	33	14	18	14	14	13
35-49 Years	14	31	12	15	36	26	36	35	24	24	26	20	15	22	15
50-64 Years	21	30	19	23	26	38	17	22	19	24	18	5	10	13	4
65 +	1	0	1	1	2	1	5	3	2	1	2	0	0	1	0

### Intestinal Transplants per Year

	2014	2013	2012	2011	2010	2009	2008	2007	2006	2005	2004	2003	2002	2001	2000
< 1 Year	3	1	9	8	7	22	29	37	31	26	23	10	15	20	10
1-5 Years	17	25	33	22	37	56	49	56	50	45	53	45	30	35	30
6-10 Years	20	26	42	30	44	78	78	93	81	71	76	55	45	55	40
11-17 Years	2	5	6	3	8	5	5	10	9	10	6	11	13	3	3
18-34 Years	9	12	18	13	25	21	34	27	34	33	14	18	14	14	13
35-49 Years	14	31	12	15	36	26	36	35	24	24	26	20	15	22	15
50-64 Years	21	30	19	23	26	38	17	22	19	24	18	5	10	13	4
65 +	44	73	49	51	87	85	87	84	77	81	58	43	39	49	32

## End of Transplant?

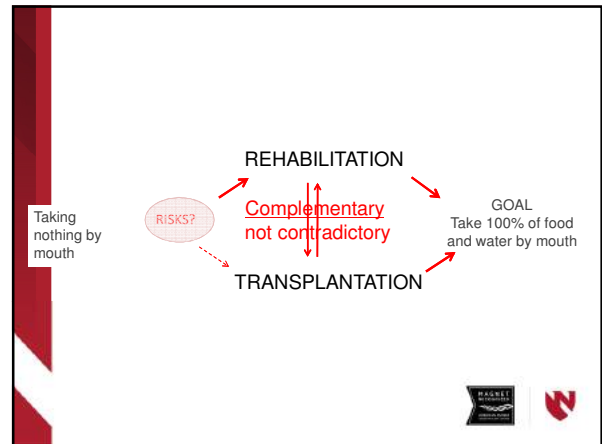

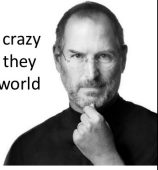
- Liver Disease**
  - Omegaven/SMOF has improved liver condition/function
  - Risk of GI bleeding from varices minimized improved liver function
- Central Line Infections**
  - Ethanol Lock solutions have minimized central line infections
  - Yeast and gram negative organisms falling – these were the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

# 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?

