Intestinal Failure
(Introduction)

Iradj Maleki MD
Mazandaran University of Medical Sciences
iradj2001@yahoo.com
Content of lecture

- Definition of intestinal failure (IF)
- Classifications of IF
- Historical aspect of IF
- Etiologies of IF in children
- Etiologies of IF in adults
- General approach to IF
Definition of intestinal failure (IF)

- A clinical condition of temporary or permanent intestinal malabsorption

- Reduction in functioning gut mass below the minimal amount necessary for adequate digestion and absorption of nutrients
Enteral nutrition support

Enteral feeding for hundreds of years was enema (eggs, wine, milk, whey and wheat broth)

1958: hollow tube attached to a bladder – direct into esophagus

Later: soft, flexible leather tube into stomach
Development of parenteral nutrition

Galenic belief: circulation of blood came from liver

1628: Harvey showed the circulation in vessels came from and to the heart

First IV injection of ethanol in dog showed the same effects as oral ingestion

Later: Cholera was treated with an electrolyte and milk injection

IV lipid injection killed the dogs rapidly
Development of parenteral nutrition

1800: subcutaneous lipid injection (painful!)

Late 1800s: first infusions of glucose and amino acids in man

1924: continuous glucose infusion
**Historical aspect**

- **Newer era in the management of IF**
- Until 1960s malnutrition due to IF was fatal
- 1967: first intestinal transplant reported (died)
- 1970: successful TPN was developed
- Cyclosporine did not improve the result of intestinal transplant (ITx)
- 1990: introduction of tacrolimus changed the success rate of ITx
Intestinal failure in children
Intestinal failure in children

- Natural history of IF is influenced by underlying cause
- The causes range from congenital defects to severe and permanent disorders of early life
- Short bowel syndrome (SBS) is the first recognized cause
- But other causes have been known and described recently
Causes of IF in children

- Short Bowel Syndrome

- Intestinal neuromuscular motility disorders
  - Extended Hirschsprung’s disease
  - Chronic intestinal pseudo-obstruction
  - Congenital disease of enterocyte development
Hirschsprung’s disease

- In Hirschsprung’s disease aganglionic segment is confined to rectum & sigmoid in 80% of cases.
- Incidence of Hirschsprung’s disease: 1/5000.
- In 1% of them aganglionic section involves entire small intestine.
- No surgical procedure for improvement.
- Dependency to long-term PN is high.
Chronic intestinal pseudo-obstruction

- Heterogeneous conditions
- Myopathic or neuropathic
- Variable presentation, histopathology, severity in motility disorder, and outcome
- Urinary tract may be also involved
- Intestinal transplantation becomes a logical option
Congenital diseases of enterocyte development

- Microvillus atrophy
- Intestinal epithelial dysplasia (tufting enteropathy)

Both are inherited as autosomal recessive

Onset within days/weeks of birth with watery diarrhea

Require permanent PN

Logical candidates for intestinal transplantation
The most common cause of IF in children
Characterized by malabsorption status after extensive loss or resection of small bowel
Anatomical & functional definition
Nowadays parenteral nutrition allows infants and children to have normal growth during the long period of adaptation of small intestine
**Etiology**

- Small intestinal atresia (multiple)
- Gastroschisis (5% chance)
- Postnatal necrotizing enterocolitis (resection of ileum and proximal colon)
- Extensive Hirschsprung's disease (colon and small intestine – 50% of ileum)
- Midgut volvulus

**Preterm vs. fullterm infant!**

- Small bowel doubles during the last trimester
Short Bowel Syndrome in children

- Prognosis depends on:
  - Extension of resection/loss
  - Site of resection/loss

- Jejunal resection is tolerated better than ileum
- Capacity of adaptation is limited in jejunum
- Ileum is the site of intestinal hormones (enteroglucagon)
Short Bowel Syndrome in children

Goals of treatment are:
- Promote gut adaptation
- Recover intestinal autonomy
- Nutritional support for normal growth & development

Until 30 yrs ago prognosis for these children was poor
Now more than 90% of infants and children survive after extensive small intestinal resection with the use of daily parenteral & enteral nutrition
Duration of PN depends on:
- Length of intestine
- Presence of ileo-cecal valve

Other prognostic factors:
- Functional capacity of intestine (absorption & motility)
- Bacterial Overgrowth (BOG)
Short Bowel Syndrome in children

- Improvement of functional capacity (surgical)
  - Longitudinal intestinal lengthening and tailoring (LILT) (1980)
  - Serial transverse enteroplasty (STEP) (2003)

- Management of BOG:
  - Antibiotic
  - Probiotic (?)
  - Surgical corrections
Intestinal failure in adults
Etiologies of intestinal failure in adults

- Iatrogenic
  - Blunt trauma
  - Penetrating trauma
  - Operative misadventure

- Ischemic
  - Arterial embolism
  - Venous occlusion
  - Low flow/shock

- Obstructive
  - Adhesive
  - Internal hernia
  - Radiation enteritis

- Infiltrative
  - Desmoid tumors
  - Carcinoid
  - Amyloidosis
  - Malignancy

- Functional
  - Pseudo-obstruction
  - IBD
  - Bacterial overgrowth
Short Bowel Syndrome in adults

• SBS represents nearly 80% of long-term need for home parenteral nutrition
• In adults minimal length of a normal small intestine are:
  • 100 cm with no colon
  • 60 cm with some colon
  • 35 cm with ileo-cecal valve and full colon
Causes of SBS in adults are:

- Ischemia (21%)
- Crohn's disease (16%)
- Desmoid tumor, Gardner's syndrome and familial polyposis (24%)
- Trauma (12%)
- Volvulus (9%)
- Other (radiation enteritis, postbariatric surgery, ...) (20%)
Short Bowel Syndrome in adults

- SBS in Crohn’s diseases is due to
  - Extensive primary disease
  - Multiple resections
  - When considering intestinal transplantation, recurrence of primary disease must be considered!

- SBS due to venous thrombosis may be due to inherited hyper-coagulable disorders like:
  - Factor V Leyden mutation
  - Protein C or S deficiency
  - Anti-thrombin deficiency
Intestinal failure related to bariatric surgery

Procedure types:

- Jejuno-ileal bypass
- Bilio-pancreatic diversion
- Roux-en-Y gastric bypass
- Distal gastric bypass
Intestinal failure related to bariatric surgery

- Bariatric procedures can lead to IF
- They are at risk for SBS due to post-op complications
  - Internal hernia
  - Mesenteric ischemia
  - Bypassed segment
- Reversion of procedure may be necessary!
- Liver transplant may be necessary!
SBS due to IBD (Crohn's disease)

- SBS is due to fistulas, obstruction, abscess, perforation in CD and mesenteric vein thrombosis in CD or ulcerative colitis
- Management of SBS is like other etiologies
- Newer biologic therapies has reduced the need for resections, and hence the prevalence of SBS
- Nutritional deficiencies should be addressed and treated (vitamins B12, A, D, E, K, iron, selenium, Zn, ...)

SBS is due to fistulas, obstruction, abscess, perforation in CD and mesenteric vein thrombosis in CD or ulcerative colitis.

Management of SBS is like other etiologies.

Newer biologic therapies has reduced the need for resections, and hence the prevalence of SBS.

Nutritional deficiencies should be addressed and treated (vitamins B12, A, D, E, K, iron, selenium, Zn, ...).
General approach to management of Intestinal failure
Goals of management of IF

- Cure the IF (weaning PN)
- Produce a safe artificial support
- Offer the best quality of life with or without intestinal transplant

Patients are to be defined as candidates for
- Rehabilitation (temporary IF)
- Long-term and definite PN (irreversible IF)
- Transplantation (irreversible IF with complications)
Rehabilitation

Is considered for any patient intolerant for oral feeding

Wean in children:
- 20 cm of jejuno-ileum with ileo-cecal valve (ICV) present
- 40 cm of jejuno-ileum without ICV

Wean in adults:
- End jejunostomy (no colon) (100 cm)
- Jejuno-coli (60 cm)
- Jejuno-ileal (full colon present) (35 cm)
Probability of weaning is less than 10% if weaning is not obtained:

- In the first 4 years in children
- In the first 2 years in adults

Patients with no chance of intestinal rehabilitation are candidates for long-term PN
Long-term Parenteral Nutrition (PN)

- Minimal input of water, minerals, protein, energy
- Insufficient oral intake may compromise oral autonomy may induce higher rate of PN dependency
- Avoidance of complications:
  - Liver disease
  - Sepsis
Complication of long-term PN

- IF associated liver disease occurs in 40-60% of infants requiring long-term PN and in 15-40% of adults on HPN
- Progression to biliary cirrhosis and portal hypertension occurs in a minority
- Sepsis is the major cause of death
- If complications occurs, intestinal transplantation should be considered
**Intestinal transplantation (ITx)**

- **Indications:**
  - Failure of PN
  - Liver failure
  - Thrombosis of at least 2 major vessel channels
  - Frequent line infections (> 2 sepsis/ year)
  - Non-reconstructible GI tract
  - Difficult electrolyte management

- Late referral for ITx → higher mortality rates
Summary

- IF, the reduction of sufficient functioning gut mass can occur in all age groups
- SBS is the major etiologic group for IF in all ages
- The natural history of IF and SBS depends on many factors, including the etiology and the extension of resected and retained gut
- Management includes rehabilitation and nutritional support with PN, until weaning can be practiced
1- پیش آگهی کدامیک از انواع نارسایی روده ای بهتر است؟
الف) زژنوستومی
ب) زژنوستومی
پ) فر کی ندارد
د) فردی ندارد

c) زژنوم- آناناستوموز
ج) زژنوم- آناناستوموز

2- محدودیت در کدام ماده غذایی در نارسایی روده ای ناشی از طیف از زژنوم- آناناستوموز ضروری می باشد؟
الف) چربی
ب) چربی
پ) پروتئین
د) پروتئین

الف) زیرینه
ب) زیرینه
پ) زیرینه
د) زیرینه

3- در اختلال آب و الکترولیت ناشی از دفع بالای زژنوم- آناناستوموز که به تجویز خوراکی محلول قند و نمک- داروها پیش از کنترل پدیده می‌باشد؟
الف) اختلاص مینرالوکورتیکوئید
ب) اختلاص مینرالوکورتیکوئید
پ) اختلاص مینرالوکورتیکوئید
د) اختلاص مینرالوکورتیکوئید

الف) اختلاص مینرالوکورتیکوئید
ب) اختلاص مینرالوکورتیکوئید
پ) اختلاص مینرالوکورتیکوئید
د) اختلاص مینرالوکورتیکوئید

4- در نارسایی روده ای شدید کدام را در نظر می‌گیرید؟
الف) Parentral Nutrition
ب) Enteral Nutrition
پ) Transplantation
د) Oral Nutrition

الف) Parentral Nutrition
ب) Enteral Nutrition
پ) Transplantation
د) Oral Nutrition

5- مهم‌ترین عامل در تعیین وابستگی دراز مدت به تغذیه وریدی کدام است؟
الف) طول باقی مانده زژنوم
ب) وجود زژنوم
پ) بیماری التهابی در روده
د) بیماری التهابی در روده

الف) طول باقی مانده زژنوم
ب) وجود زژنوم
پ) بیماری التهابی در روده
د) بیماری التهابی در روده