

# Nephrotic Syndrome in Children

A scenic view of a coastal town built on a hillside, overlooking a turquoise bay with large rocks in the foreground. The town is densely packed with buildings, and the bay is a vibrant blue-green color. The sky is a clear, deep blue. The foreground is dominated by large, smooth, light-colored rocks.

# Objectives

- Define Nephrotic Syndrome
- Pathogenesis of Oedema
- Causes of Nephrotic syndrome in Children
- Minimal Change disease

# Nephrotic Syndrome

Histopathological process

OR

Clinical Entity

# Nephrotic Syndrome

Histopathological process

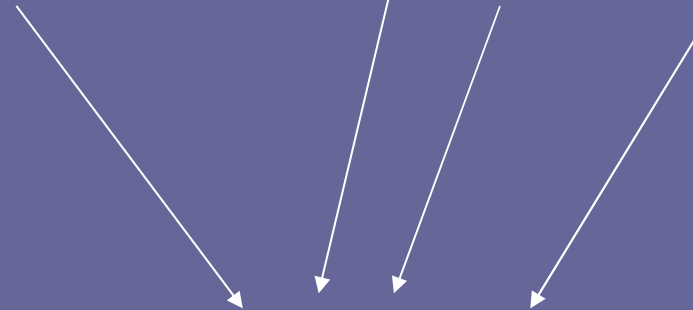
OR

Clinical Entity 😊

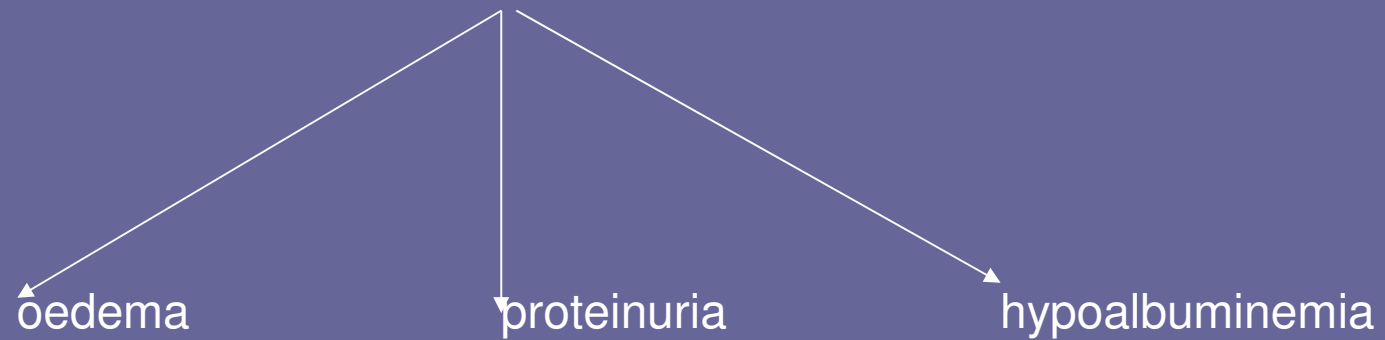
# Nephrotic Syndrome

- Massive proteinuria  $> 1 \text{ g/m}^2/\text{day}$ 
  - Spot prot:creat ratio  $0,2 \text{ g/mmol}$
- Hypoalbuminaemia
- Oedema
- Hyperlipidaemia

Primary glomerulopathies AI post infective vasculitides



**Nephrotic syndrome  
(clinical entity)**



Biopsy: glomerulonephritis

# Oedema: Pathogenesis

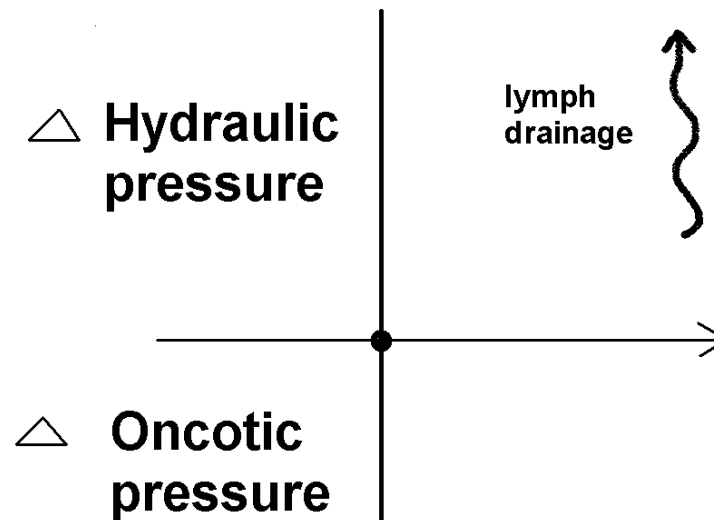
# Pathophysiology of oedema

Two basic steps:

- Alteration of capillary haemodynamics that favours movement of fluid from vascular space to interstitium

- +/- Retention of Na and H<sub>2</sub>O by the kidneys

## Water movement







Macroscopic Haematuria

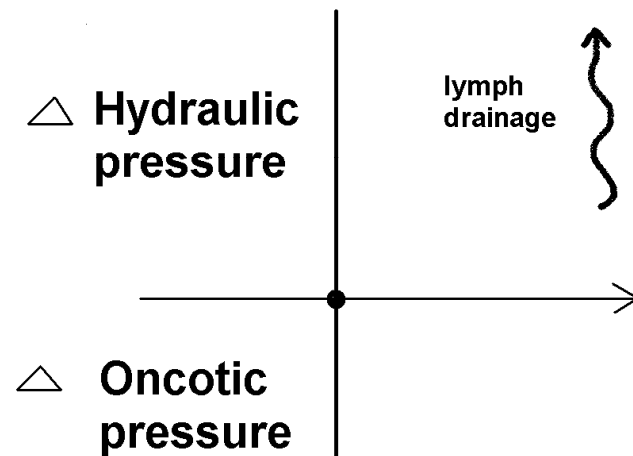
Hypertension

Generalized oedema

Creat and Urea increased



## Water movement



- Not sick looking  
5yr M

- 1 week onset

  - puffy eyes

  - swelling legs

- Urine +++ protein

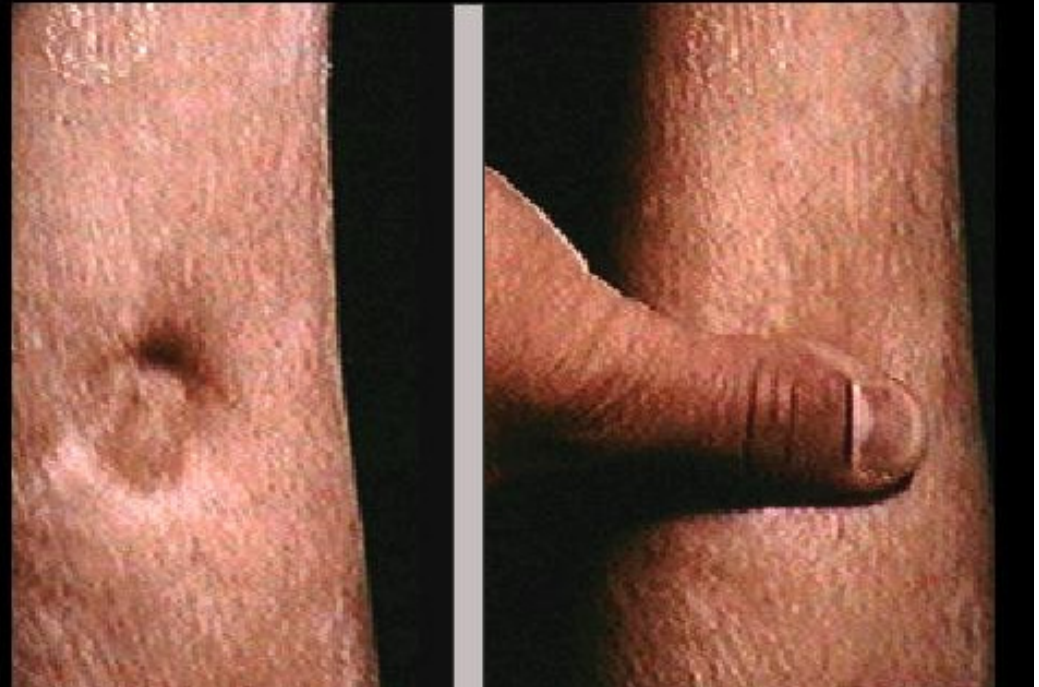
- Lab

  - alb 16

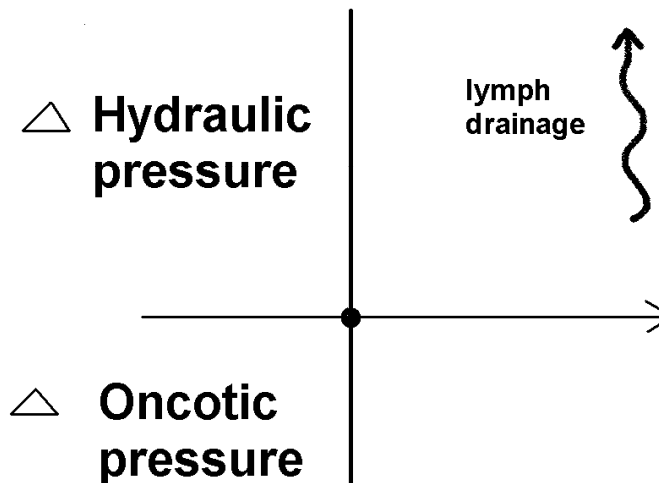
  - cholesterol 8

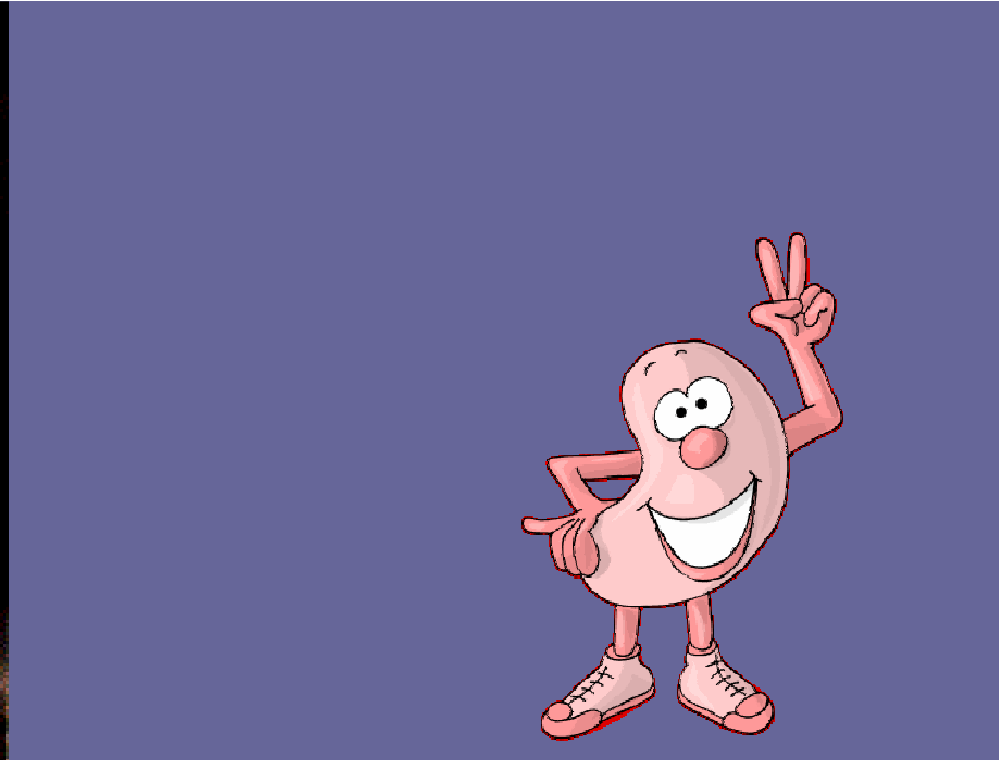
  - U+E Normal

- 

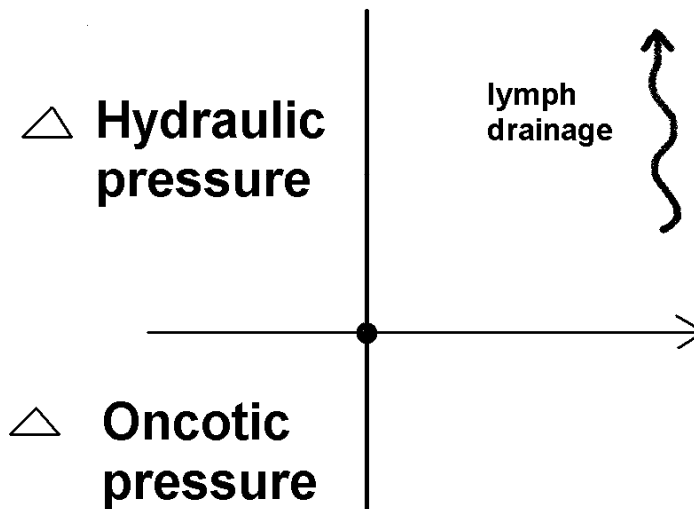


## Water movement



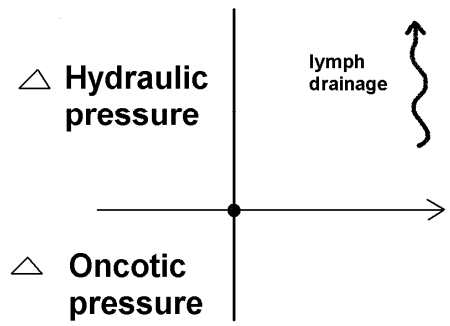


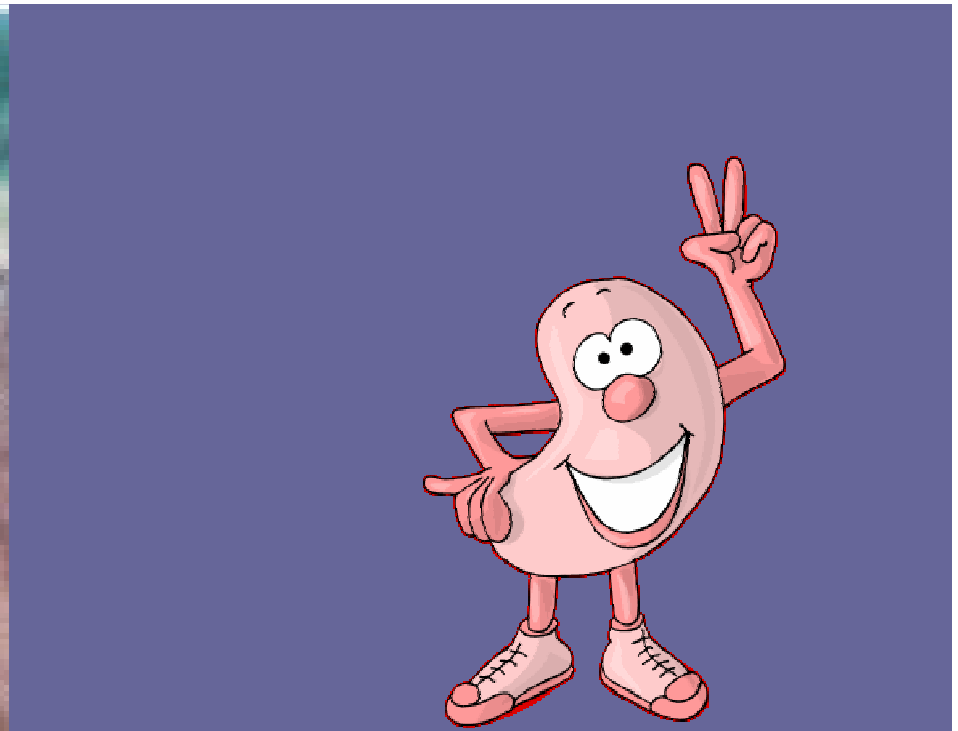
## Water movement



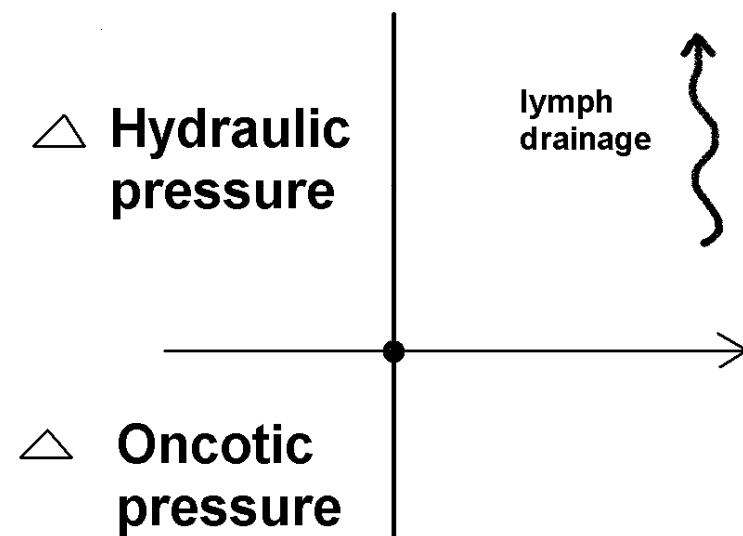


### Water movement

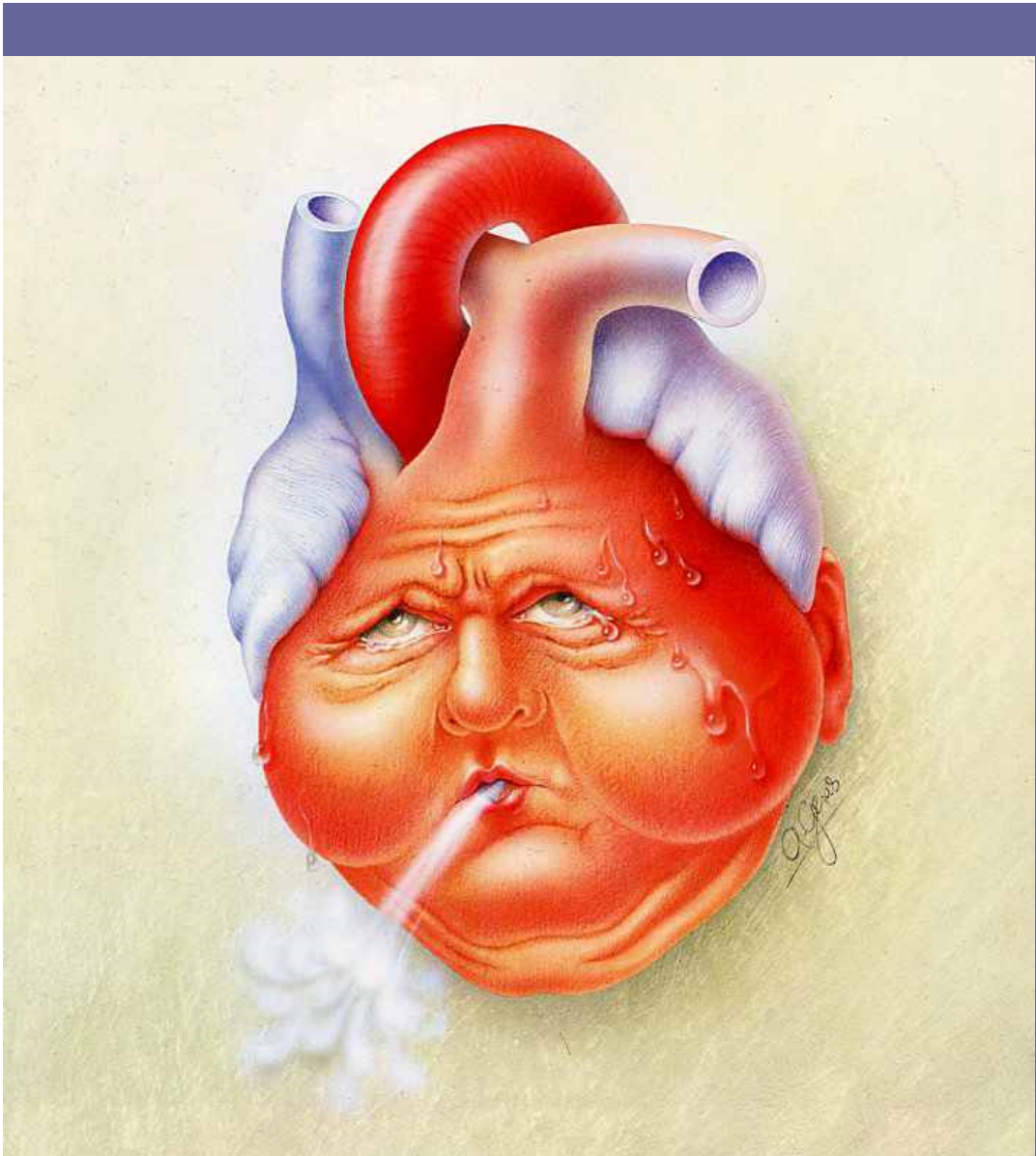




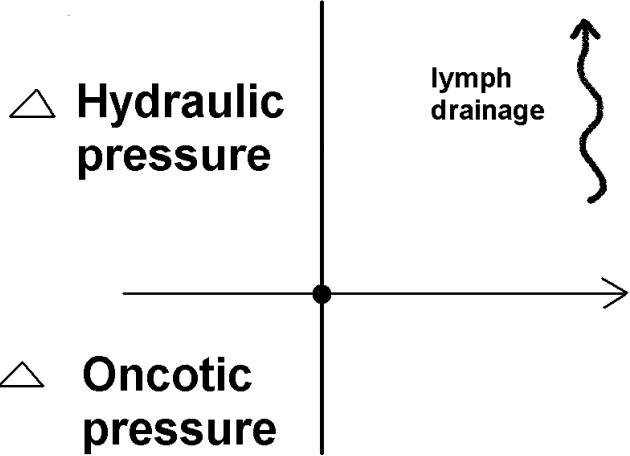
## Water movement

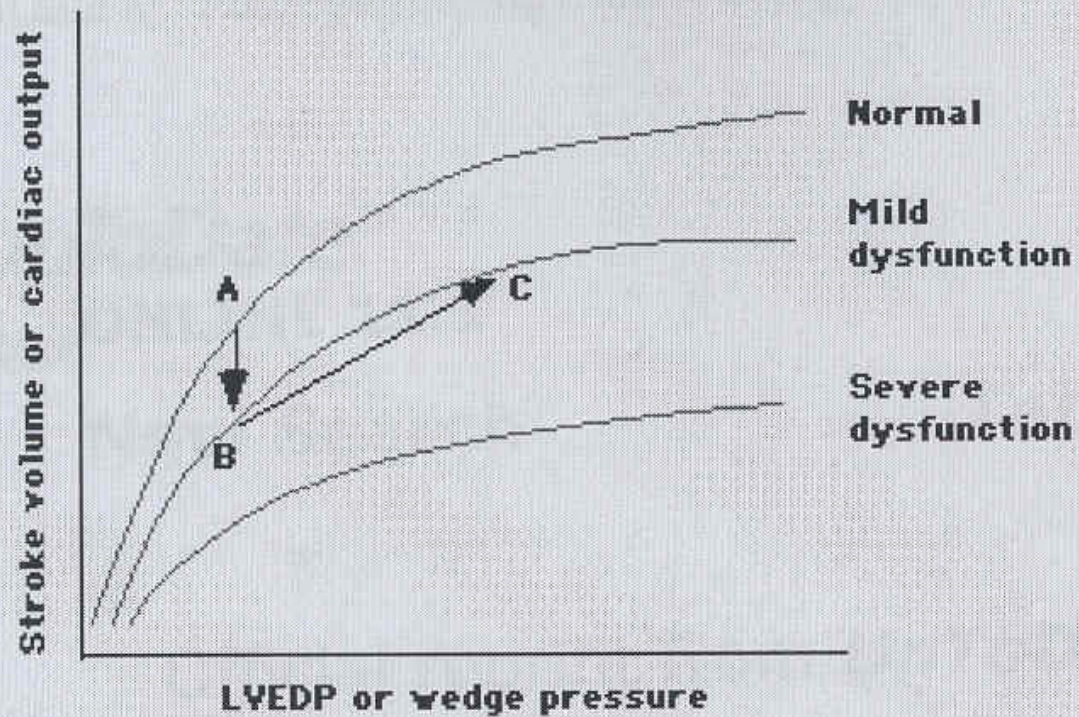




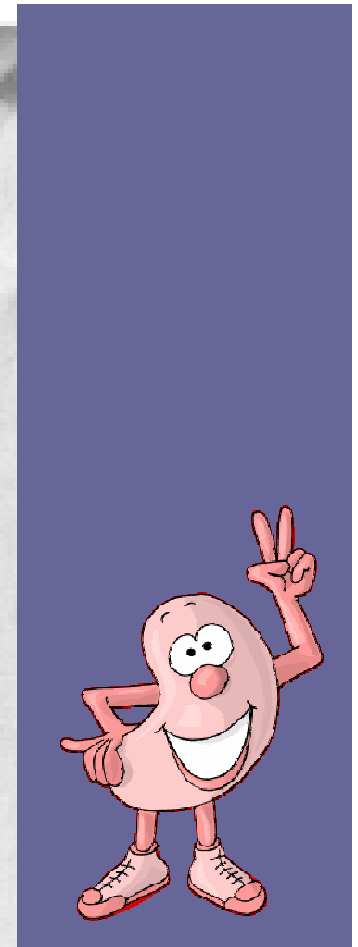
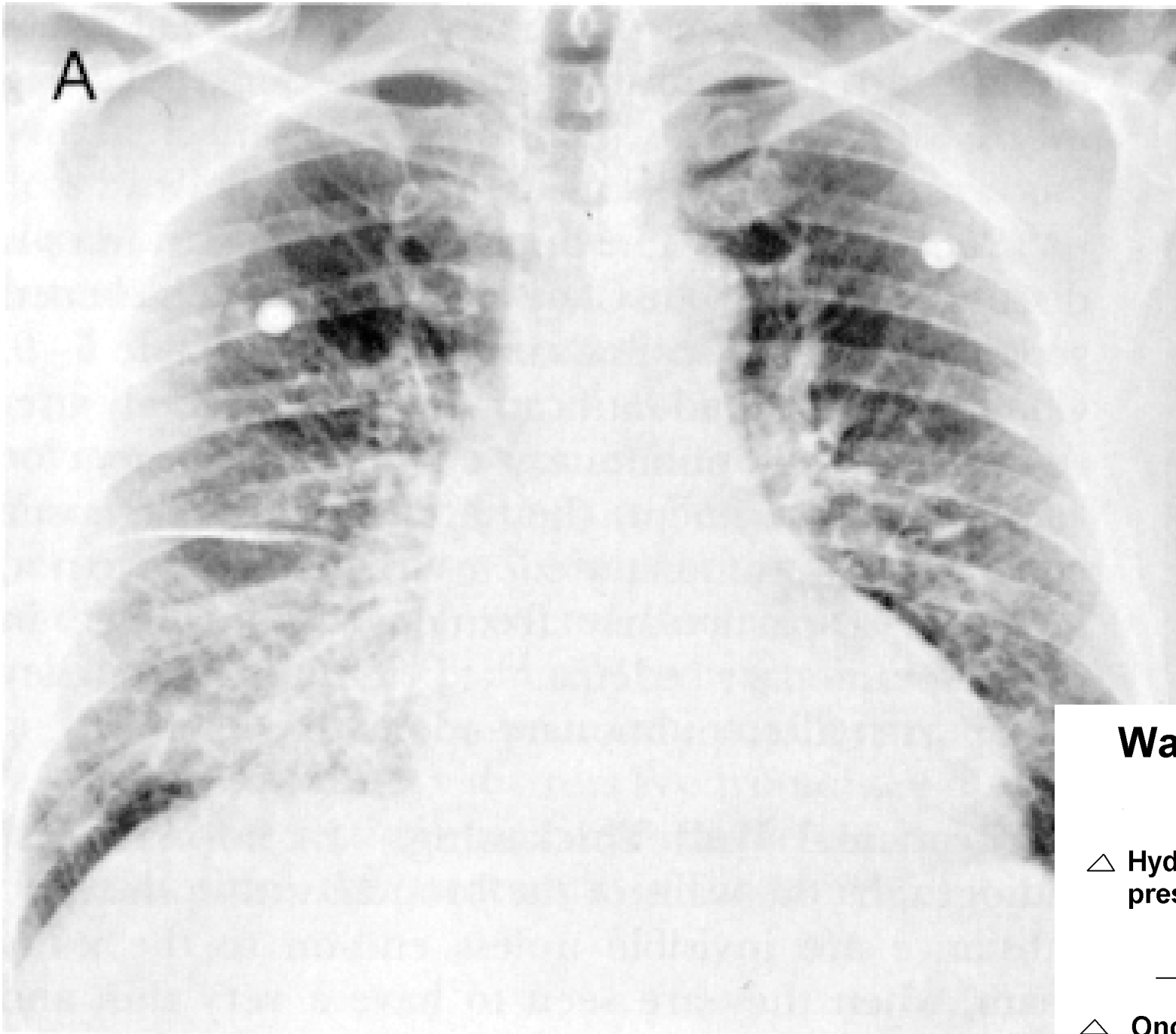


# Water movement

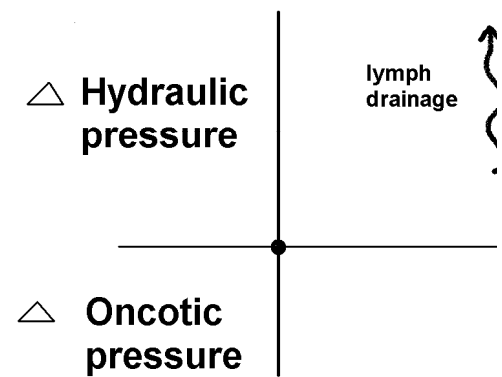




**Frank-Starling curves in CHF** Idealized family of Frank-Starling curves



## Water movemen





# Causes of Nephrotic Syndrome @ RXH

- Minimal change 43%
- Mesangioproliferative 17%
- Membranous 16%
- APSGN 6%
- MCGN 2%
- HIV ?

# Causes of Nephrotic Syndrome in black children(Gauteng)

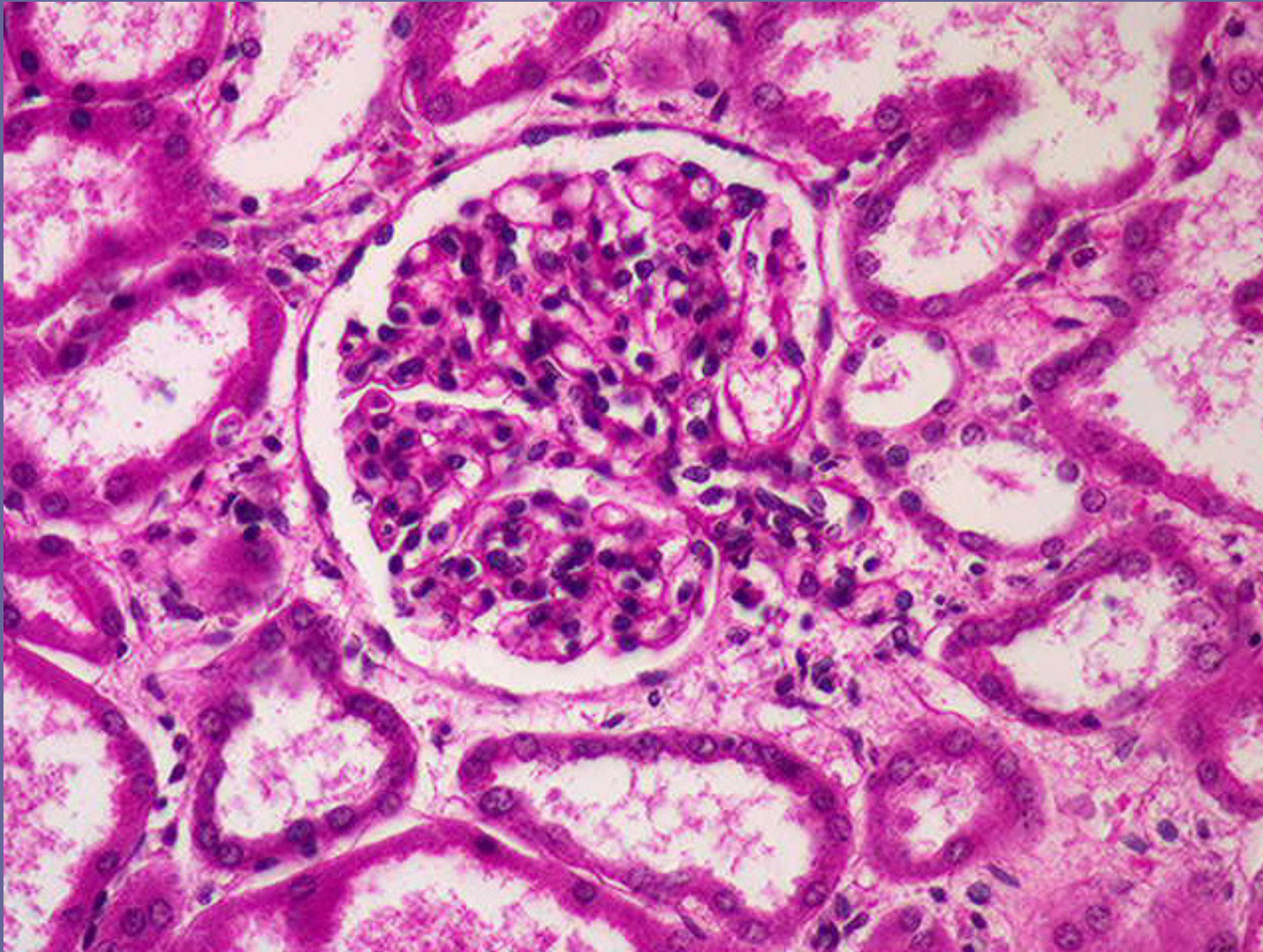
- FSGS 31%
- Minimal change 24%
- Membranous 13%
- Mesangial proliferative 13%
- Mesangiocapillary 4%

*Thomson et al*

# Pathogenesis of Minimal change Disease

- T cell dysfunction with release of lymphokines
  - Podocyte dysfunction
  - Loss of electronegative charge of basement membrane
- Light microscopy no changes
- Immunofluorescence negative
- Electron microscopy fusion foot processes

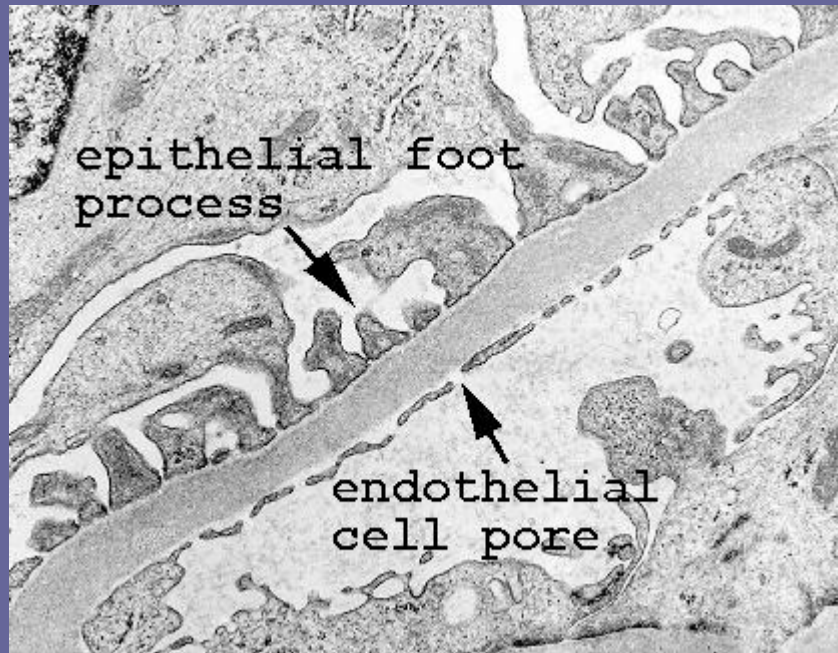
# Light microscopy : Minimal change disease



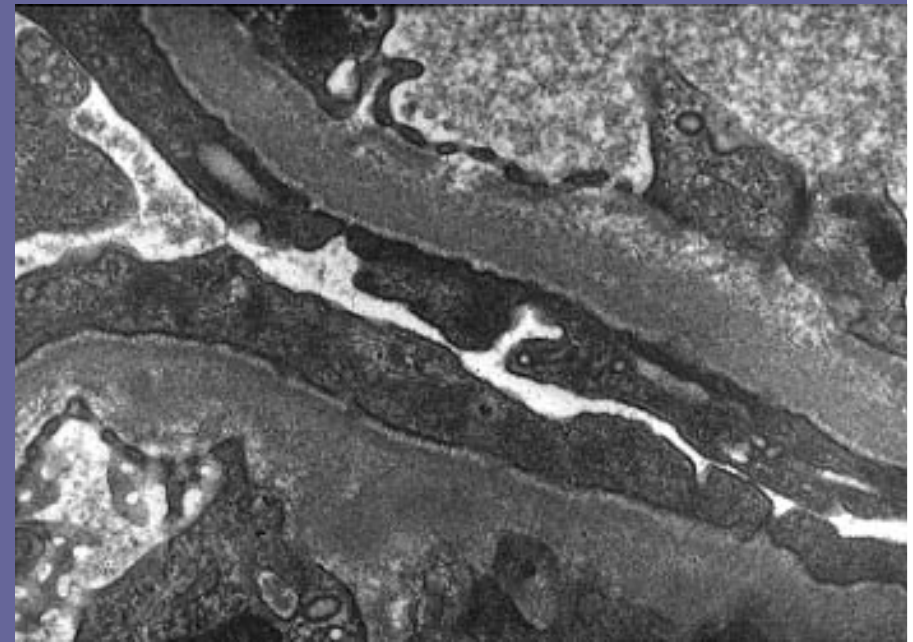


# Electron Microscopy

Normal



Fusion of foot processes



# Clinical presentation

- Rapid onset of oedema progresses rapidly
- Transudation into body cavities
  - Pleural effusion
  - Ascites
- Eyelids swell totally closed
- Severe scrotal/vulval oedema
- Urine may become frothy

# Nephrotic Syndrome Complications

- Hypovolaemic crisis
- Pre –renal failure
- Thrombosis
- Ascites
  - Hernia umbilical inguinal
- Malnutrition
- Infection
  - Peritonitis, pneumonia, cellulitis
- Hyperlipidaemia
  - Cardiovascular risk
- Hypothyroidism

# Nephrotic Syndrome

## Investigations

- Quantify how severe nephrotic syndrome?
  - quantify proteinuria
  - Serum alb cholesterol
- Are there any underlying diseases that could be causing this?
  - Urine microscopy: (bland urine in MCNS)
  - Hepatitis B, VDRL, ASOT/ANtiDNAse B, ANA, C3
- Is pt hypovolaemic
  - Clinical signs of shock
  - Urine Na
- Is renal function normal (some diseases may cause renal failure)
  - Creat Urea
- Probably going to use immunosuppressive treatment :
  - Exclude infection
  - Exclude TB



# Indications for biopsy

Indications that it might not be Minimal Change disease:

- Age < 1 year
- Age > 10 years
- Renal Failure
- Persistent hypertension
- Macroscopic haematuria
- Microscopic haematuria(persistent)
- Evidence of other disease e.g. SLE, HSP Hep B
- Failed trial of steroids
- Black kids

# Nephrotic Syndrome Management(1)

- Daily urine dipsticks
- Salt restriction
- No fluid restriction unless hypertensive or in renal failure
- Treat oedema if severe :
  - ivi albumin and diuretics
  - Furosemide(beware of powerful diuretics on their own in acute setting)
  - Furosemide +amiloride

# Manage complications:

- Treat shock 20% Alb, plasma
- Prophylactic Aspirin
- Aggressive treatment of infection
- ACE for proteinuria (long term)
- Lipid lowering agents (long term)

# Management(2): Treat Underlying cause

- If Minimal change suspected:
  - Trial of Steroids
  - **2mg/kg/day for 4 weeks**
- If NO response → Biopsy

# Nephrotic Syndrome (minimal Change) Management

- If response with 1<sup>st</sup> episode:
  - Continue for total of 3 months steroids
  - 6 weeks daily 2mg/kg/day then
  - 6 weeks alternate day weaning over last 2 weeks
- Subsequent episodes:
  - 2mg/kg/day **daily** until urine clear for 3 days in row
  - then **alt** days for 1 month and then rapid wean

# Outcome

- 10% never relapse
- 60% infrequent relapsers
- 30% frequent relapsers or steroid dependant
- 90% permanent remission at puberty
- Minimal risk of chronic renal failure

# FSGS

- More difficult
- High dose steroids
- Cyclophosphamide
- Cyclosporin



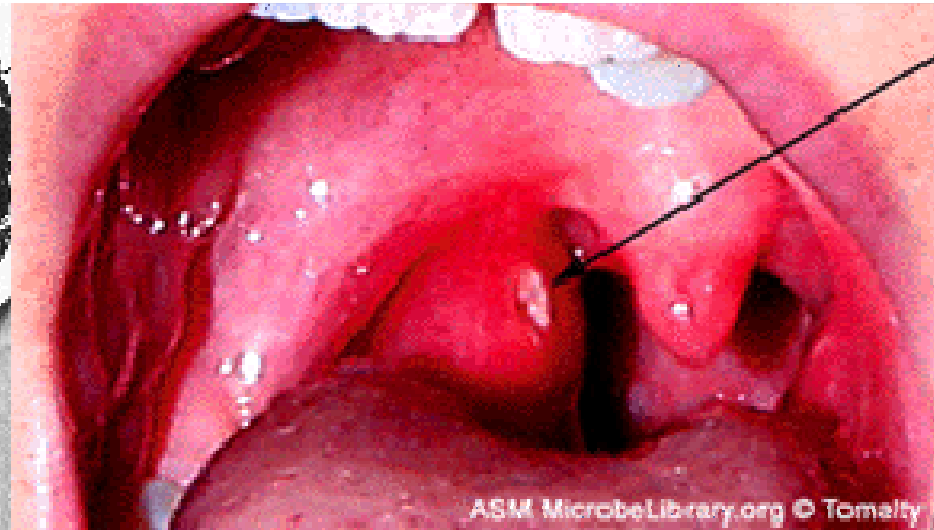


SURE, THE PLANET'S  
GOING TO GET  
A LITTLE  
WARMER.

BUT IT WON'T  
FEEL SO BAD  
'CAUSE YOU'LL BE  
ANKLE-DEEP IN  
WATER!



© 2002 Stewie



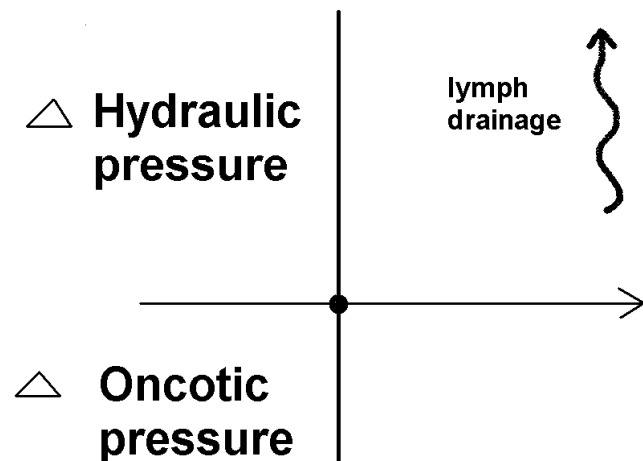
Enlarged right tonsil with adherent plaque

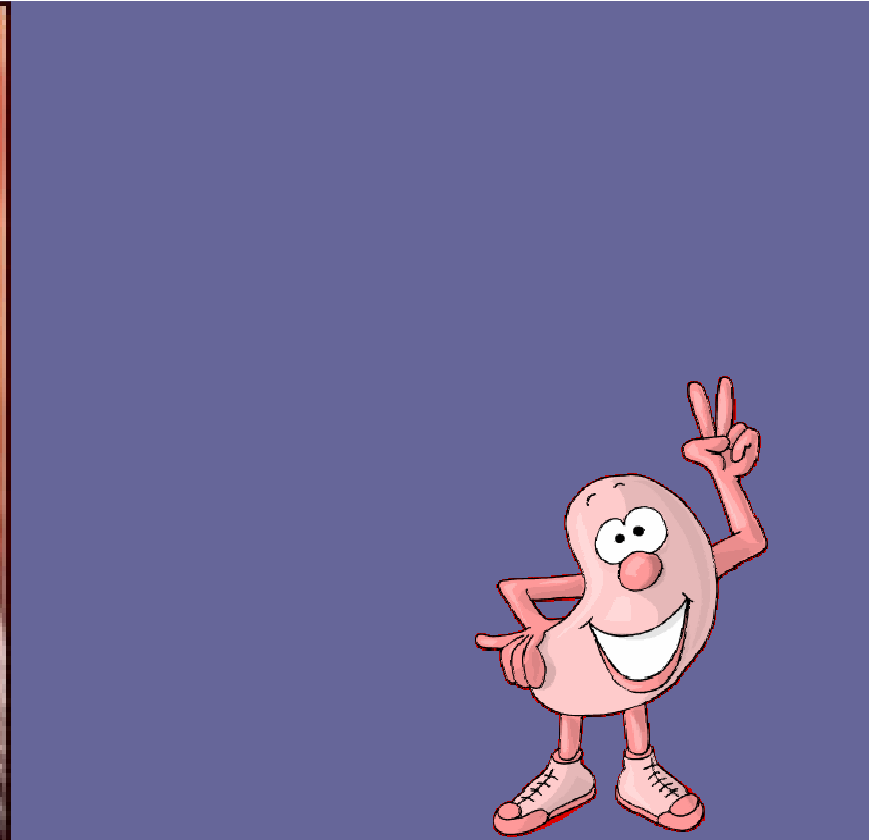
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## Water movement





## Water movement

