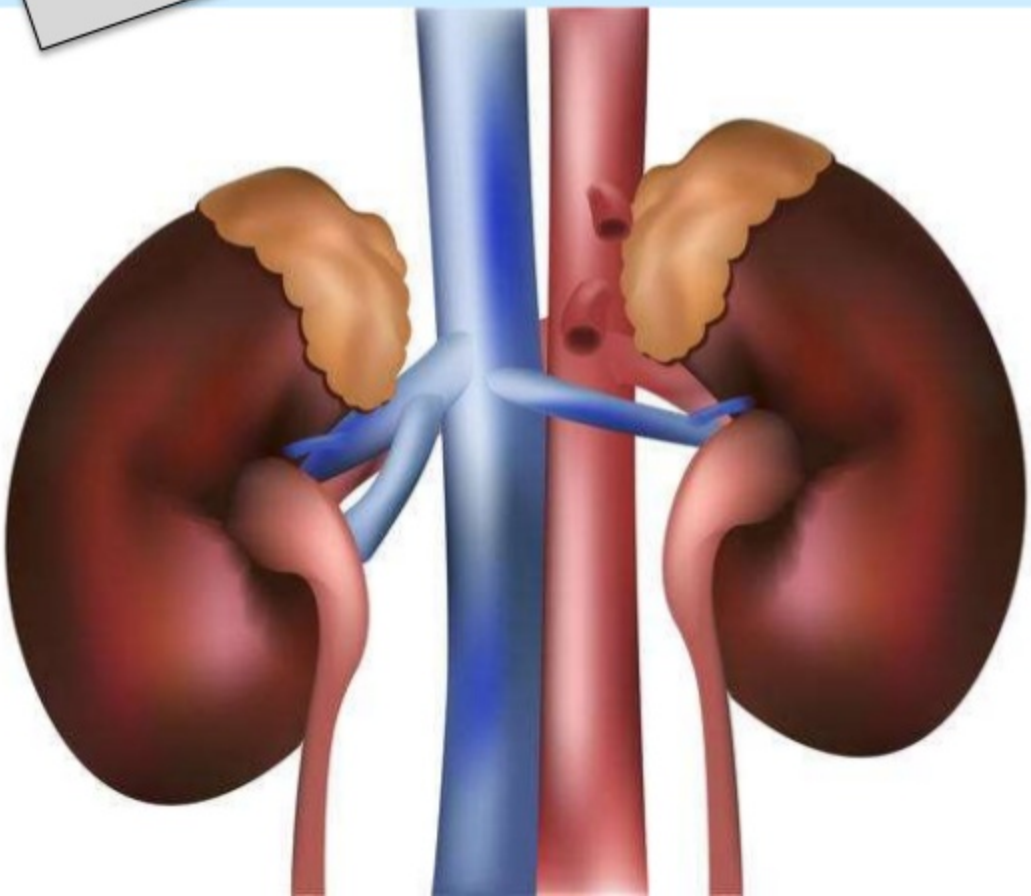
Nephrotic Syndrome



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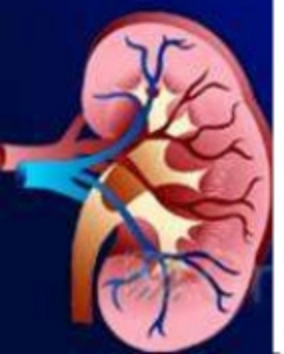
RUWI

Introduction

- Nephrotic syndrome (NS)
  - Commonest glomerular disease affecting children
  - Frequently encountered in general paediatrics
  - Characterised by
    - Significant proteinuria (early morning urine protein to creatinine ratio > 200mg/mmol) leading to
      - Hypoalbuminaemia (plasma albumin of < 25g/l)

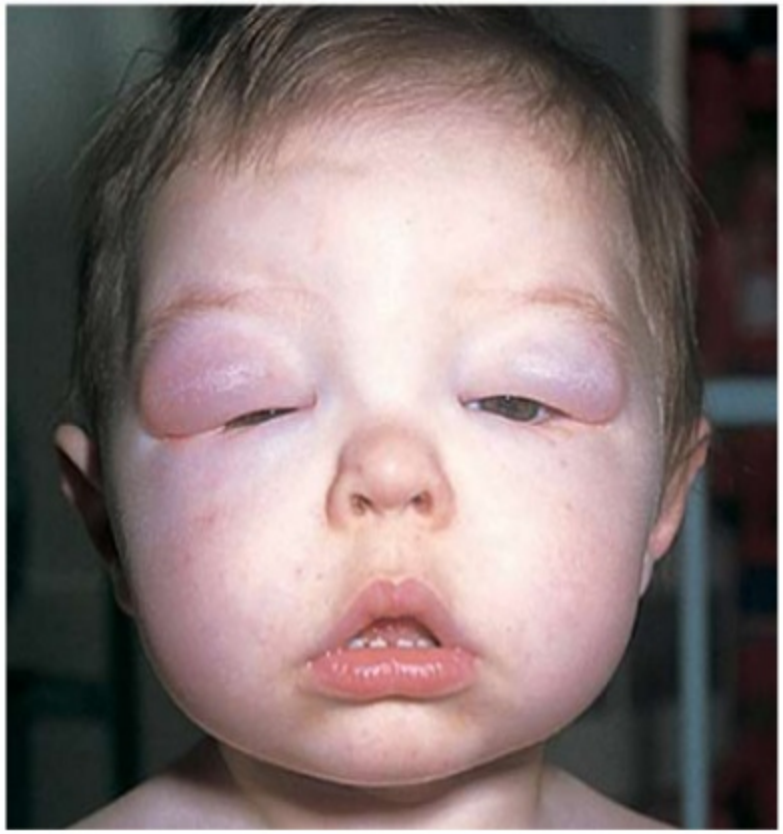


Paediatrics and child health 2010;20(1):36-42

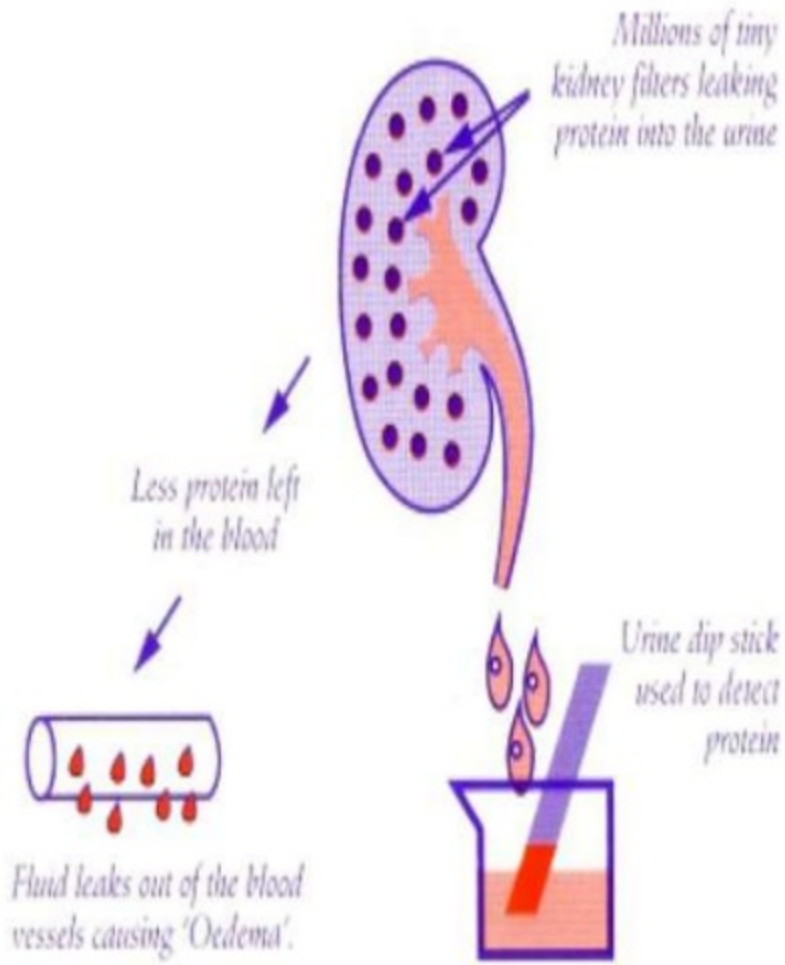
Definition

- Manifestation of **glomerular** disease, characterized by **nephrotic range** proteinuria and a triad of clinical findings associated with large urinary **losses of protein** : hypoalbuminaemia , edema and hyperlipidemia

- Nelson Textbook of Paediatrics, Vol 2, 19th Edition, page 1801

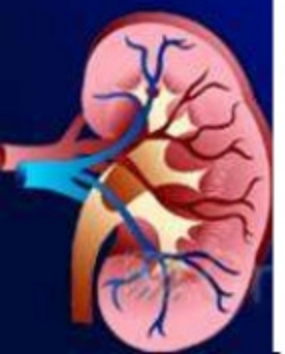


sometimes accompanied by h e m a t u r i a , hypertension and reduced glomerular filtration rate.



Why 'nephrotic range'

- Defined as
  - protein excretion of > 40 mg/m^2/hr
  - First morning protein : creatinine ratio of > 2-3 : 1



- Nelson Textbook of Paediatrics, Vol 2, 19th Edition, page 1801

Incidence (paediatric) ?

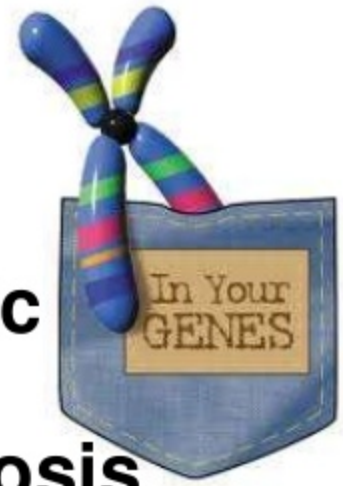
- 2 – 7 cases per 100,000 children per year
- Higher in underdeveloped countries
  (South east Asia)
- Occurs at all ages but is most prevalent in children between the ages 1.5-6 years.
- It affects more boys than girls, 2:1 ratio

http://www.kidney.org/site/107/pdf/NephroticSyndrome.pdf

Etiology

- Genetic
- Secondary
- **Idiopathic** or Primary

Genetic causes



- **Finnish type Congenital Nephrotic Syndrome**
- **Focal Segmental Glomerulosclerosis**
- **Diffuse Mesangial Sclerosis**
- **Denys-Drash Syndrome**
- **Nail – Patella Syndrome**
- **Alport Syndrome**
- **Charcot-Marie-tooth disease**
- **Cockayne syndrome**
- **Laurence-Moon-Beidl-Bardet Syndrome**
- **Galloway-Mowat Syndrome**

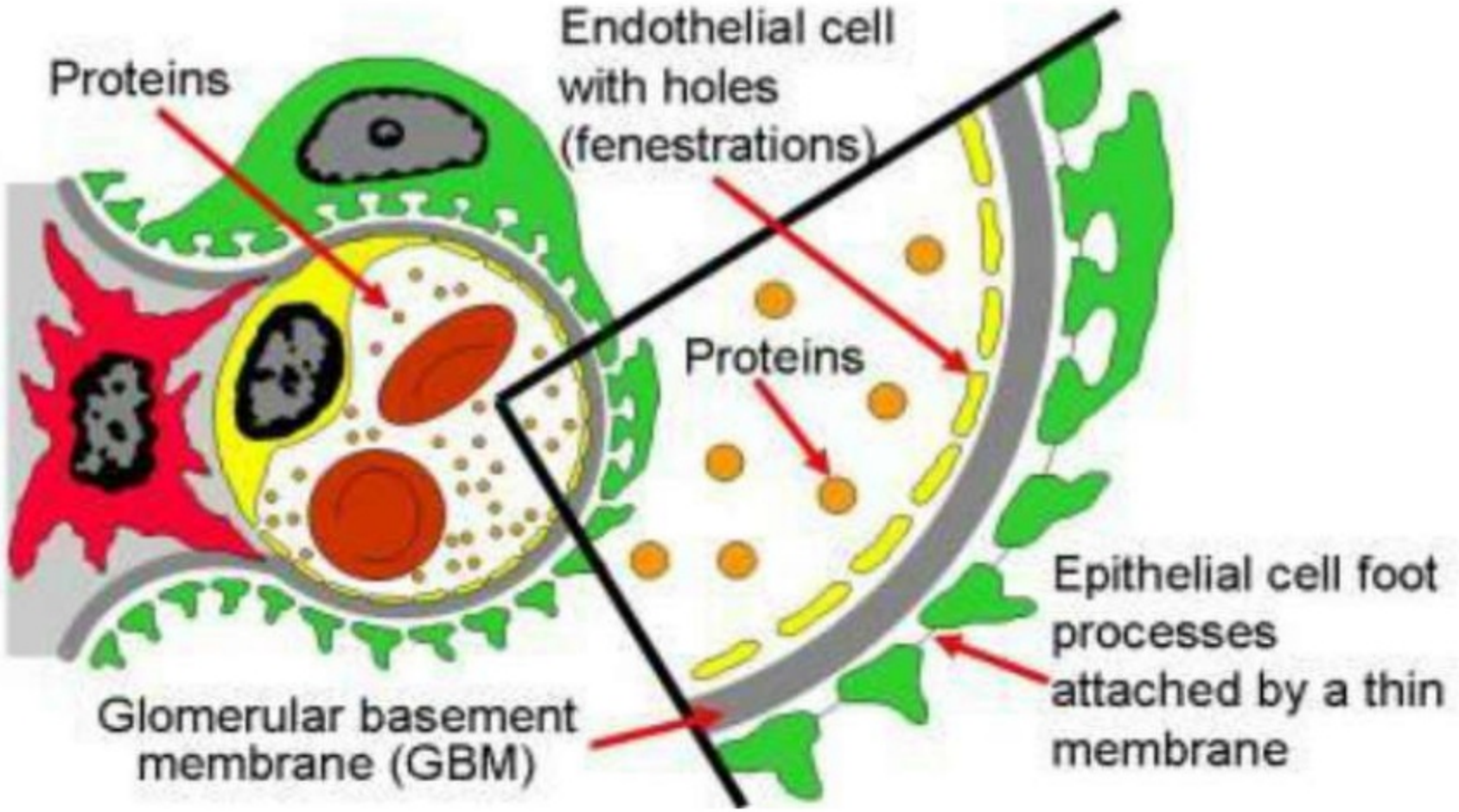
- Nelson Textbook of Paediatrics, Vol 2, 19th edition, page 1802, table 521-1

Secondary causes

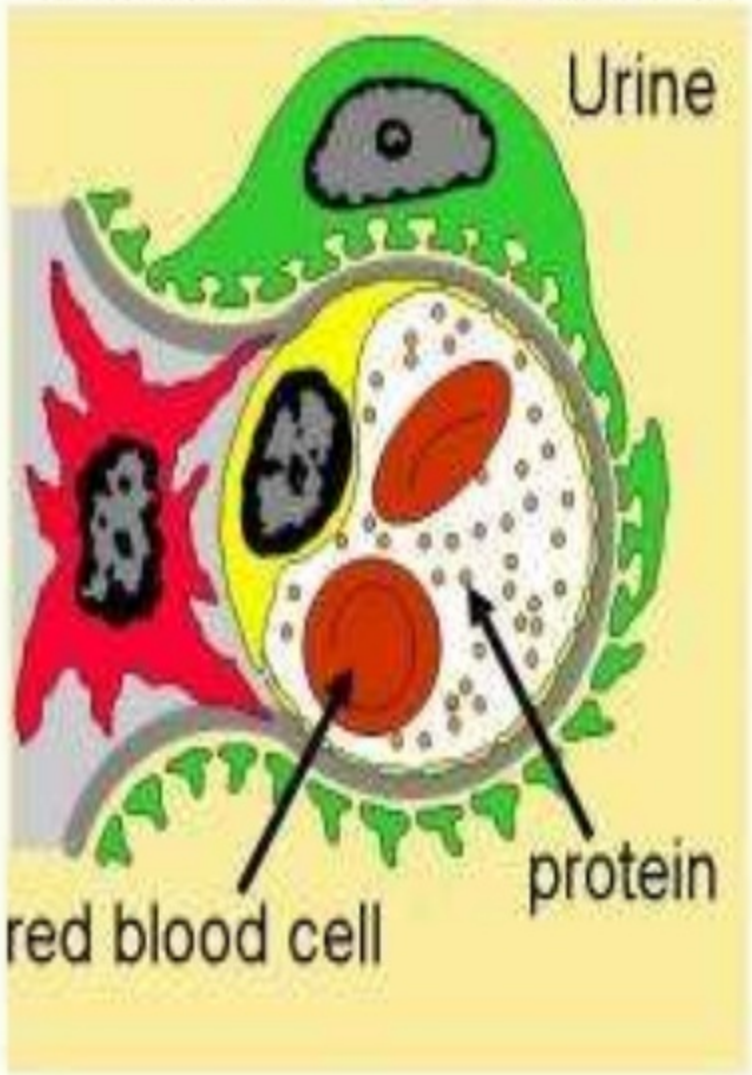
- Congenital
  - Oligomeganephronia
- Infectious
  - Hepatitis (B,C) , HIV-1, Malaria, Syphilis, Toxoplasmosis
- Inflammatory
  - Glomerulonephritis
- Immunological
  - Castleman Disease, Kimura Disease, Bee sting, Food allergens
- Neoplastic
  - Lymphoma, Leukemia
- Traumatic (Drug induced)
  - Penicillamine, Gold, NSAIDS, Pamidronate, Mercury, Lithium

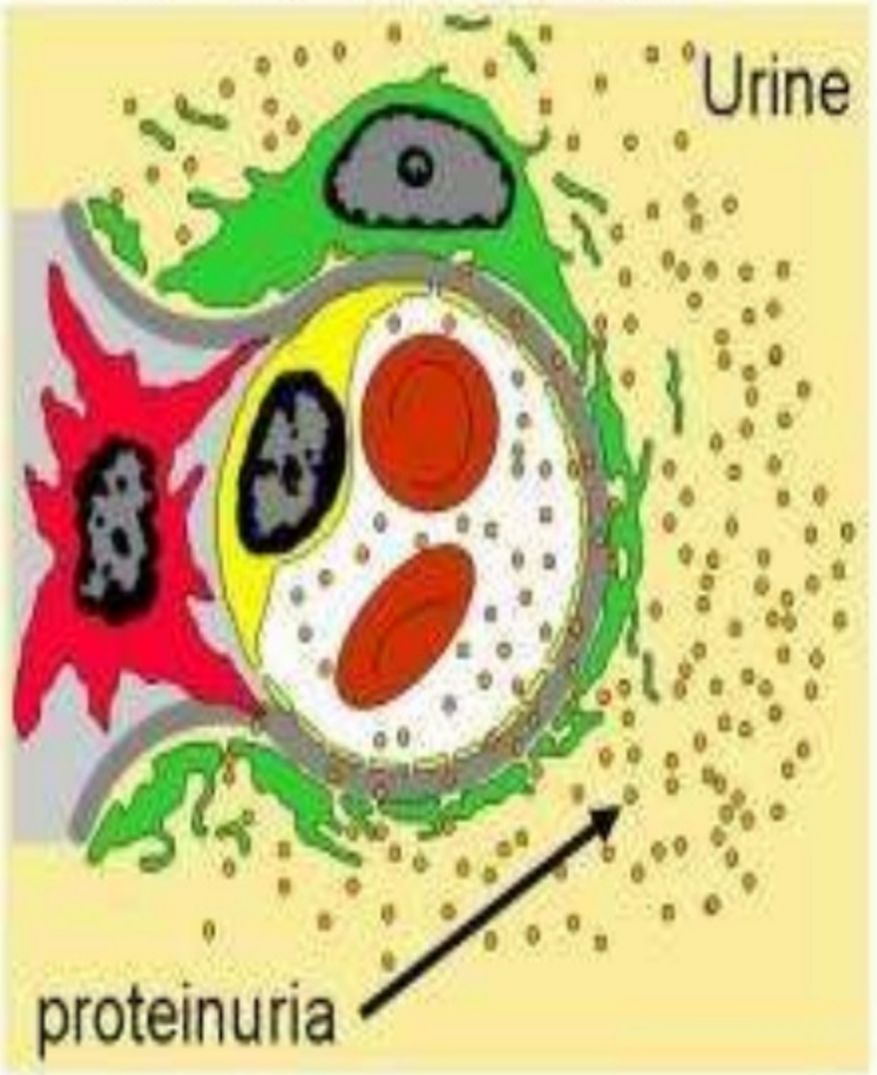
Idiopathic

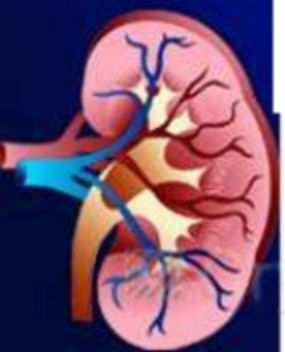
- Minimal Change disease (>80 %)
- Mesangial proliferation
- Focal segmental Glomerulosclerosis
- Membranous Nephropathy
- Membranoproliferative glomerulonephritis



Pathophysiology







Complex disturbances in immune system

Genetic Mutations / Mutations in proteins

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Extensive effacement of podocyte foot processes

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Increased permeability of the glomerular capillary wall

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Massive proteinuria

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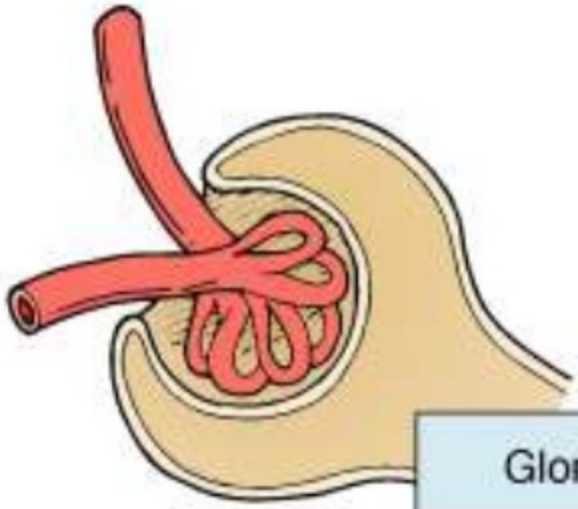
Hypoalbuminaemia

↓

Edema



Glomerular damage

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Increased permeability to proteins
Proteinuria (≥3.5 g/24 h)

↓

Hypoproteinemia

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Decreased plasma oncotic pressure

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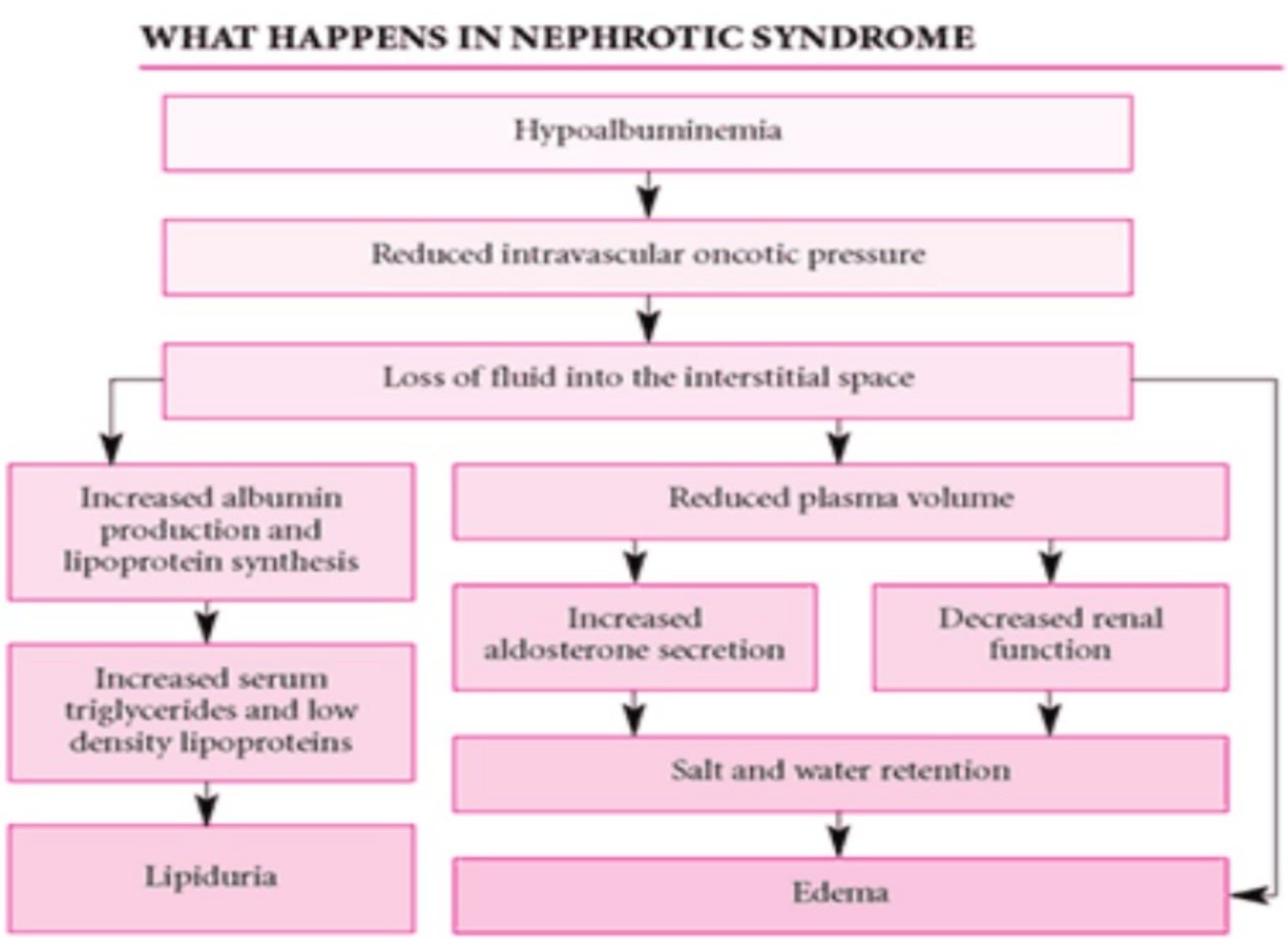
Edema

Compensatory synthesis of proteins by liver

↓

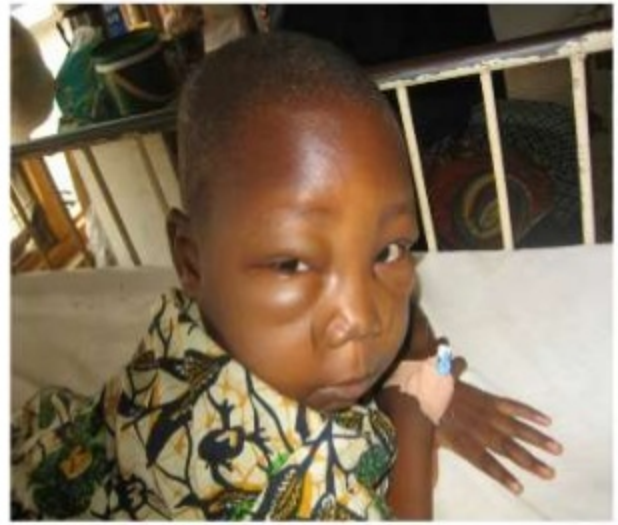
Hyperlipidemia

PATHOPHYSIOLOGY

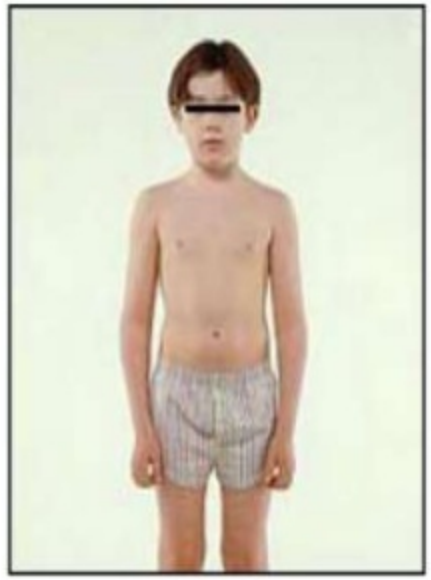


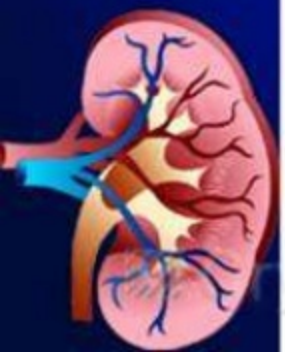
Clinical Features

CLINICAL MANIFESTATIONS



- Weight gain
- Puffiness of face (facial edema
  - -especially around the eyes
  - -apparent on rising in morning
  - -subsides during the day
- Abdominal swelling
- Pleural effusion
- Labial or scrotal swelling
- Edema of intestinal mucosa may cause:
  - diarrhea
  - anorexia
  - poor intestinal absorption



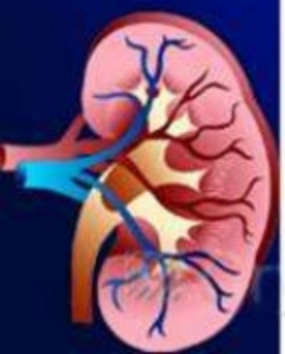


CONTD….

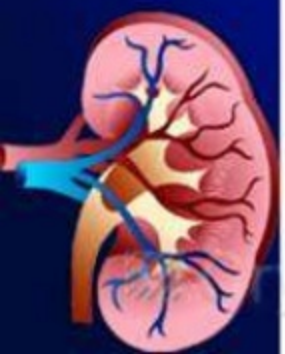
- Ankle/leg swelling
- Irritability
- Easily fatigued
- Lethargic
- Blood pressure normal or slightly decreased
- Susceptibility to infection
- Urine alterations :-

  -decreased volume

  -frothy



CLINICAL FEATURES	Minimal Change Nephrotic Syndrome	Focal Segmental Glomerulosclerosis	Membranous Nephropathy
Age (yr)	2 - 6	2 - 10	40 - 50
Sex (M : F)	2 : 1	1.3 : 1	2 : 1
Nephrotic Syndrome	100 %	90 %	80 %
Asymptomatic proteinuria	0	10 %	20 %
Hematuria	10 – 20 %	60 – 80 %	60 %
Hypertension	10 %	20 % early	infrequent
Rate of progression to renal failure	Non progressive	10 yrs	50 % in 10 – 20 yrs
Associated Conditions	Usually none	None	Renal vein thrombosis, SLE, Hepatitis B



- Nelson Textbook of Paediatrics, Vol 2 : page 1803, table 521-2