NEPHROTIC SYNDROME

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NEPHROTIC SYNDROME

OBJECTIVES

* DEFINITION&INCIDENCE
* TYPES & AETIOLOGY
* CLINICAL FEATURES
* INVESTIGATIONS
* TREATMENT & COMPLICATIONS
* PROGNOSIS

NEPHROTIC SYNDROME

Definition:-It is a clinical triad consists of:-

1)Hypoalbuminemia(serum albumin<2.5gms%).

2)Heavy proteinuria(>40mgs/m2/hr or >2gms/day)

3)Generalized edema.

4)With or without hyperlipidemia.

Incidence:-2-3/100,000. M-F ratio is 2:1

It usually affects age group from 2-8Yrs but it may start since birth(Congenital Type).

It may affect older age group.

NEPPHROTIC SYNDROME

Types of nephrotic syndrome:-

1)Congenital nephrotic syndrome;symptoms appear in the first 3-4months of life.

2)Infantile nephrotic syndrome;symptoms Starts between 4-12 months of life.

3)Idiopathic nephrotic syndrome;appear In older age group(>1Yr of age).

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Aetiology :- Depending on the cause it is divided into two types:-

1)primary(idiopathic)nephrotic syndrom(90% of cases)&it is of 3 histopathological types;

a)minimal change type(MCNS)-85%of cases and it has the best prognosis.

b)focal segmental glomerulosclrosis(FSGS)-10% of cases and it has the worse prognosis.

c)mesangioproliferative type(MP)-5%of cases.

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2)Secondary Nephrotic Syndrome(10%):-

a)Membranoproliferative GN.
b)Membranous GN.
c)Drugs(heavy metals,ACEinhibitors,NSAID)
d)Tumours(Lymphomas,Lung cancer).
e)Chronic infections(Malaria,Bilharsiasis).
f)Blood born diseases(HBV,HCV,HIV).
g)C.T Disease(SLE&Rheumatoid arthritis).

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Clinical features:-The main presentation is generalized body swelling (edema) which start as puffiness of face and lower limb swelling.

With time the swelling involve abdominal cavity (Ascites).pleural effusion&sometimes pericardial effusion may occur(ANASARCA) Genitalia may be involved.

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General symptoms like anorexia,malaise nausea,abdominal pain&irritability may be present.

Blood pressure may be normal or low due to hypovolemia.

Blood presure may be high in5-10%of cases

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Investigations:These include the followings:-

1)Urine analysis-Evidence of proteinuria of varying degrees,sometimes with hematuria.

A 24hrs urine collection to calculate total protein excrtion is mandatory for diagnosis(>40mgs/m2/hr is diagnostic of nephrotic syndrome).

Random urine sample for protein/creatinine ratio (>3)is also suggestive of nephotic syndrome.

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2)Chemistry-Total serum proteins are low

and serum albumin is <2.5gms/dl.

Serum cholesterol&triglycerides are high.

RFT(B.urea&S.creatinin) are usually normal.

Serum calcium may be low.

3)CBC TLC,Hb&Platelets.ESR may be high.

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4)Serology HBV,HCV, HIV.

5)Immunoglobulins level(IgM&IgG)

6)Complements level(C3&C4).

- Ultrasound scanning of abdomen may show edematous kidneys or sometimes an associated structural abnormalities.

- Renal biopsy is indicated in some cases.

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Treatment:-it consists of two parts:-

1)General treatment;includes the following:-

a)Diet-low salt,normal protein&low fat diet.

b)Diuretics-take care of hypotension,hypo-kalemia and renal failure.

c)Antibiotics-if with evidence of infection.

d)Salt poor albumin in some cases.

e)ACE inhibitors&ARB agents.

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2)specific treatment:It includes the following

A)Steroid therapy;oral prednisolon in a dose 60mg/m2/day as single dose for4-6 weeks then change to 40mg/m2 in alternate day as a single dose at morning for 4-6 weeks.

Tapering to be over a period of 3-6 months.

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Steroid resistant;No response after 6 weeks of daily steroid therapy.

Steroid dependent;Patient get relapse within 2 weeks of stopping steroid or changing dose.

Frequent Relapse;>2 relapses in 6 months or > 3 relapses in 1 year.

Remission is defined as absence of proteinuria for 3 cosequative days

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B)Cytotoxic therapy:-include the following,

1)Alkylating agents(Cyclophosphamide and chlorambucil).

2)Calcineurine inhibitors (Ciclosporine and tacrolimus).

3)Antipurines (Azothioprim).

4)Other imunosupresents&imunomodulators e.g mycophenolate mofetil(MMF)&levamisol.

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Vaccination which include vaccination program against pneumococci(PPV)and annual Influenza vaccine.

Prophylactic vaicella zoster immunoglobulin (VZIG)to be given to non immunized patient within 72 hours of exposure to a varicella patient .

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Complications:-

1)Infections, Peritonitis is the commonest

Others include sepsis,cellulitis,pneumonia and U.T.I

Why?

The commonest organisms are pneumococci and G-ve bacteria.

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2)Thromboembolic phenomenon(2-5%)may occur because of two factors:-

a)Increased prothrombotic factors like fibrinogen and hemoconcentration.
b)Decreased fibrinolytic factors like proteins C&S and antithrombin 3(lost in urine).

3)Renal failure(due to hypovolemia).

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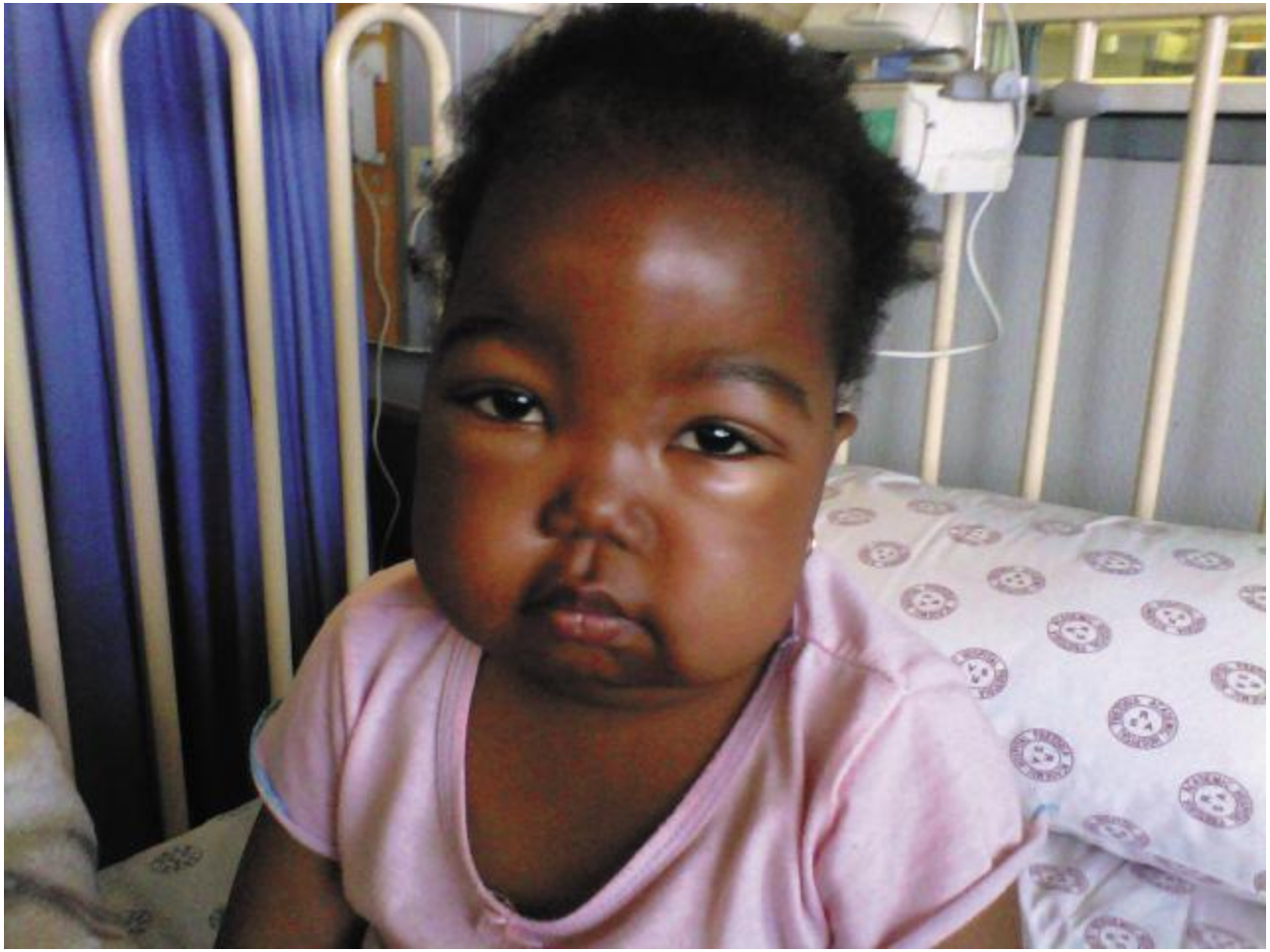
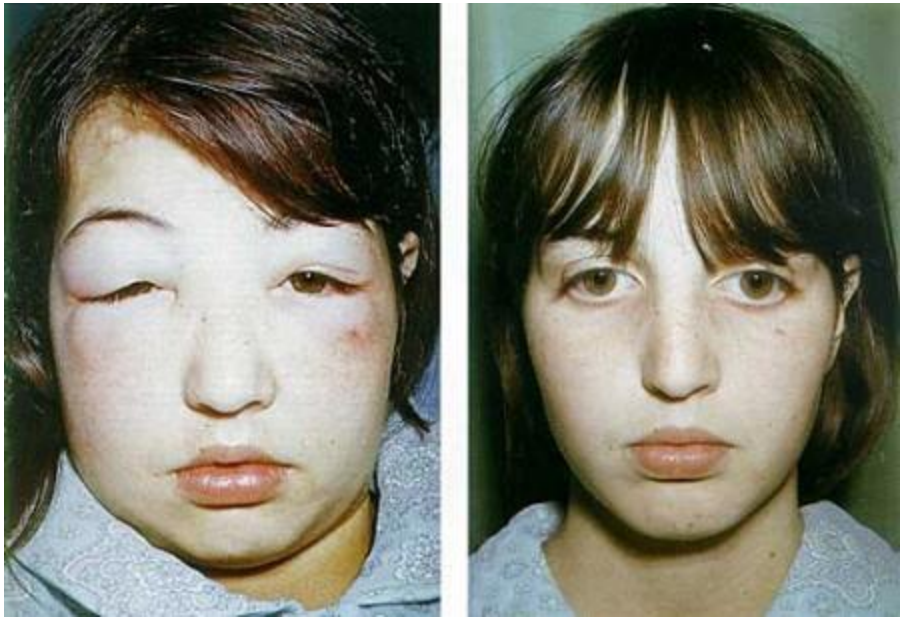
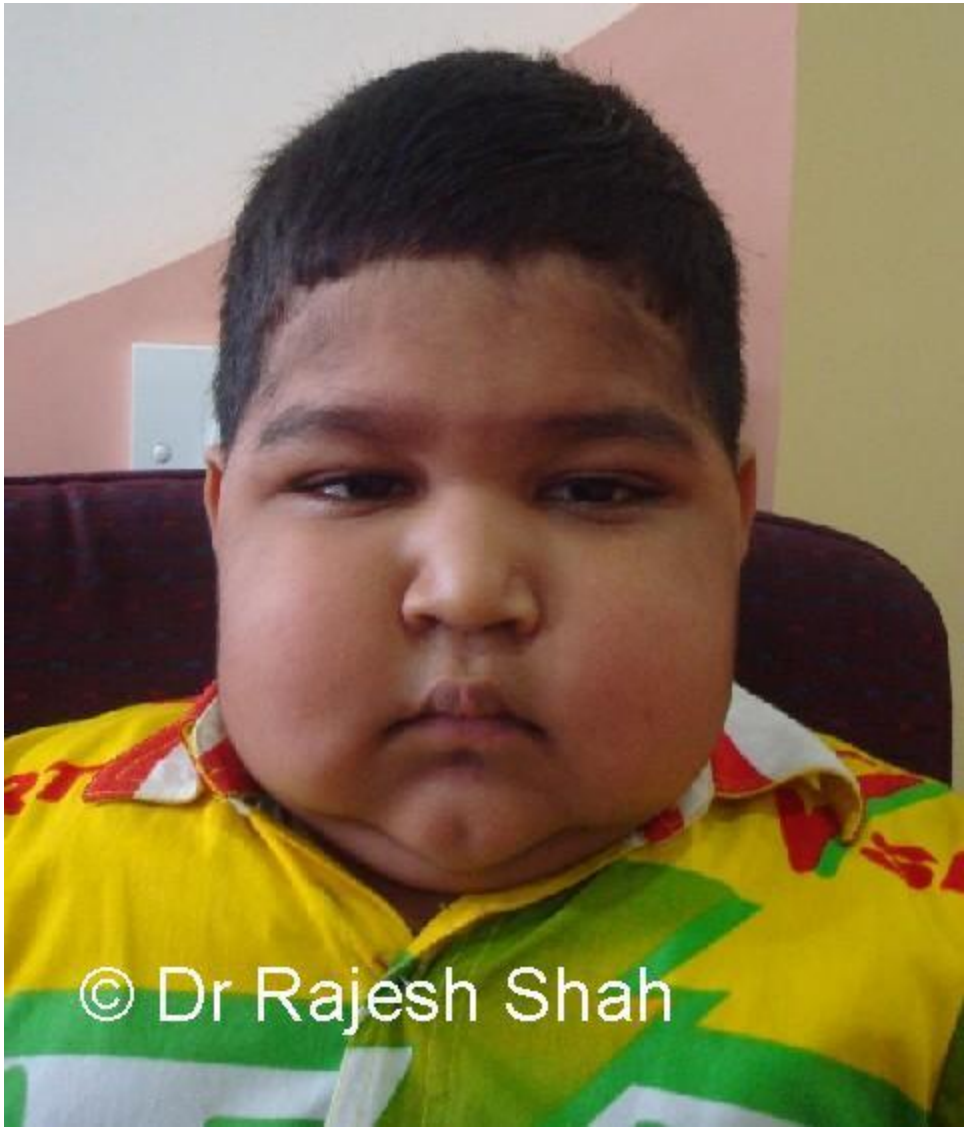
Prognosis:-More than 95% of cases(MCNS) repond to treatment &prognosis is good.
About 50% of mesengioproliferative type and only 20% of FSGS type will respond to steroid therapy.

Many patients with FSGS type will end with renal failure(ESRF)and they will need renal replacement therapy including renal transplantation.

There is a high risk of recurrence of the disease in the transplanted kidney(30-50%)in FSGS patients..



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