By,

Mr.Sachin T.Gadade

M.sc(N) Pediatrics

TERMINOLOGY:-

1. Nephroma:- Tumor of kidney

2. Nephro - Blastoma:- A rapidly developing malignant mixed tumor of kidney

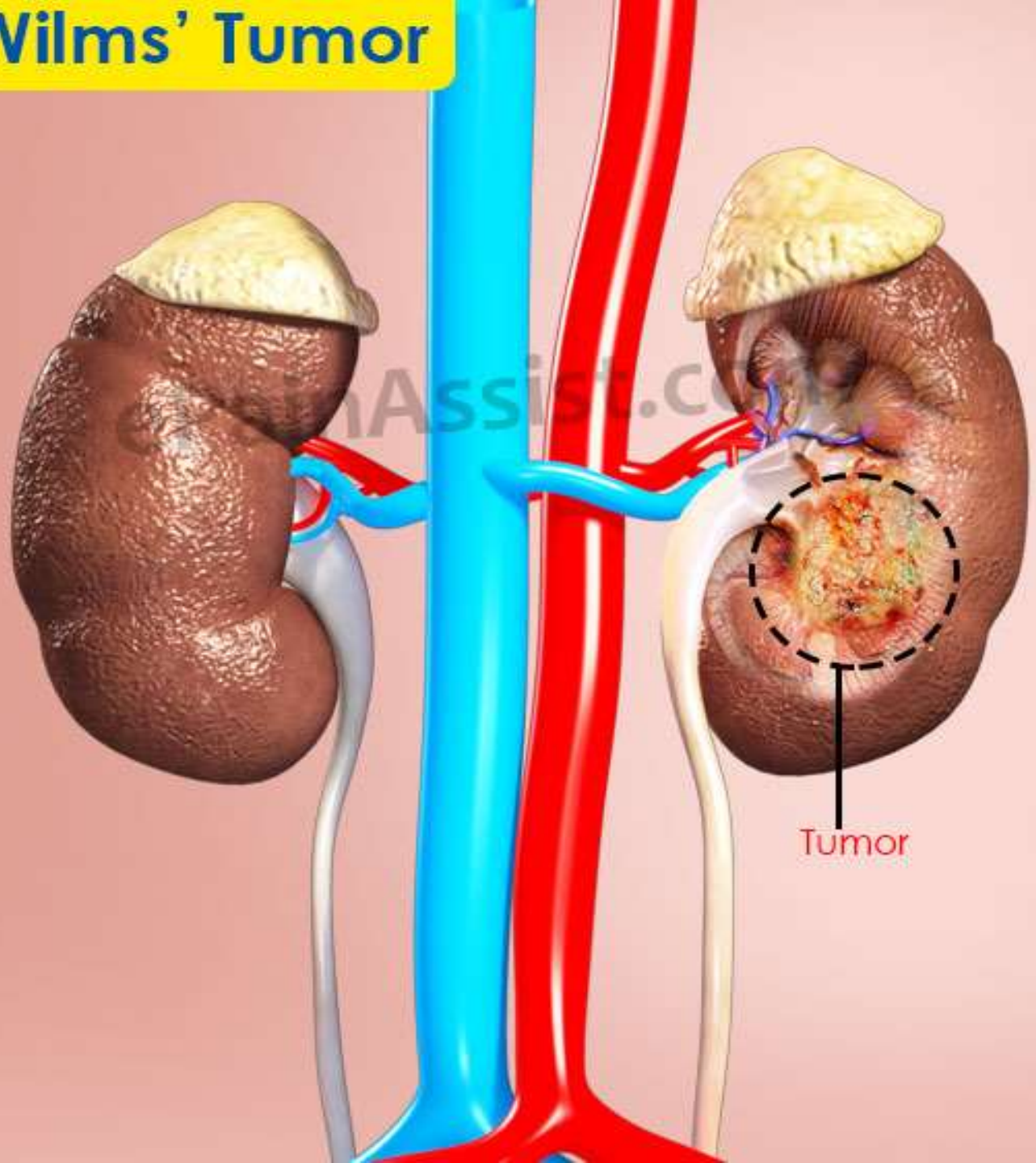
3. Primitive cells:- Premature cells

4. Clumps:- To collecting together into clumps

5. Aniridia:- Lack of part or the whole of iris

6. Renin:- An enzyme secreted by juxtaglomerular apparatus of kidneys that converts angiotensinogen to angiotensin

Wilms' Tumor



Wilms' tumor is commonly seen in kids from ages 3 to 4 and tends to occur less frequently after the age of 5. The most common kidney cancer in children is Wilms' tumor.

Signs & Symptoms of Wilms' Tumor

1) Swelling in the abdomen.
2) Pain in the abdomen.
3) Mass in the abdomen which can be felt.
4) Fever.
5) Hematuria or blood in the urine.

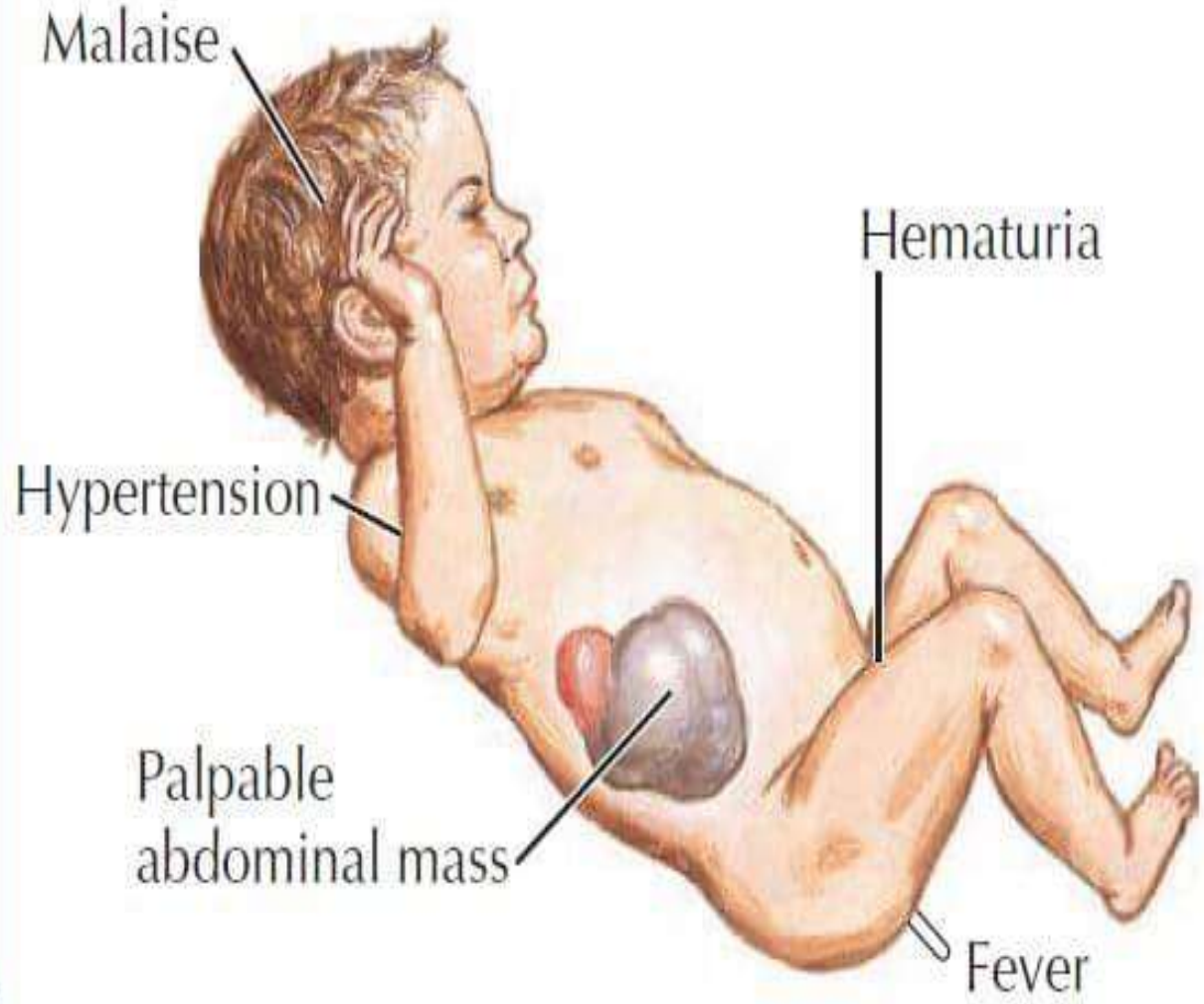
Genetics, Presentation, and Radiographic Findings of Wilms Tumor

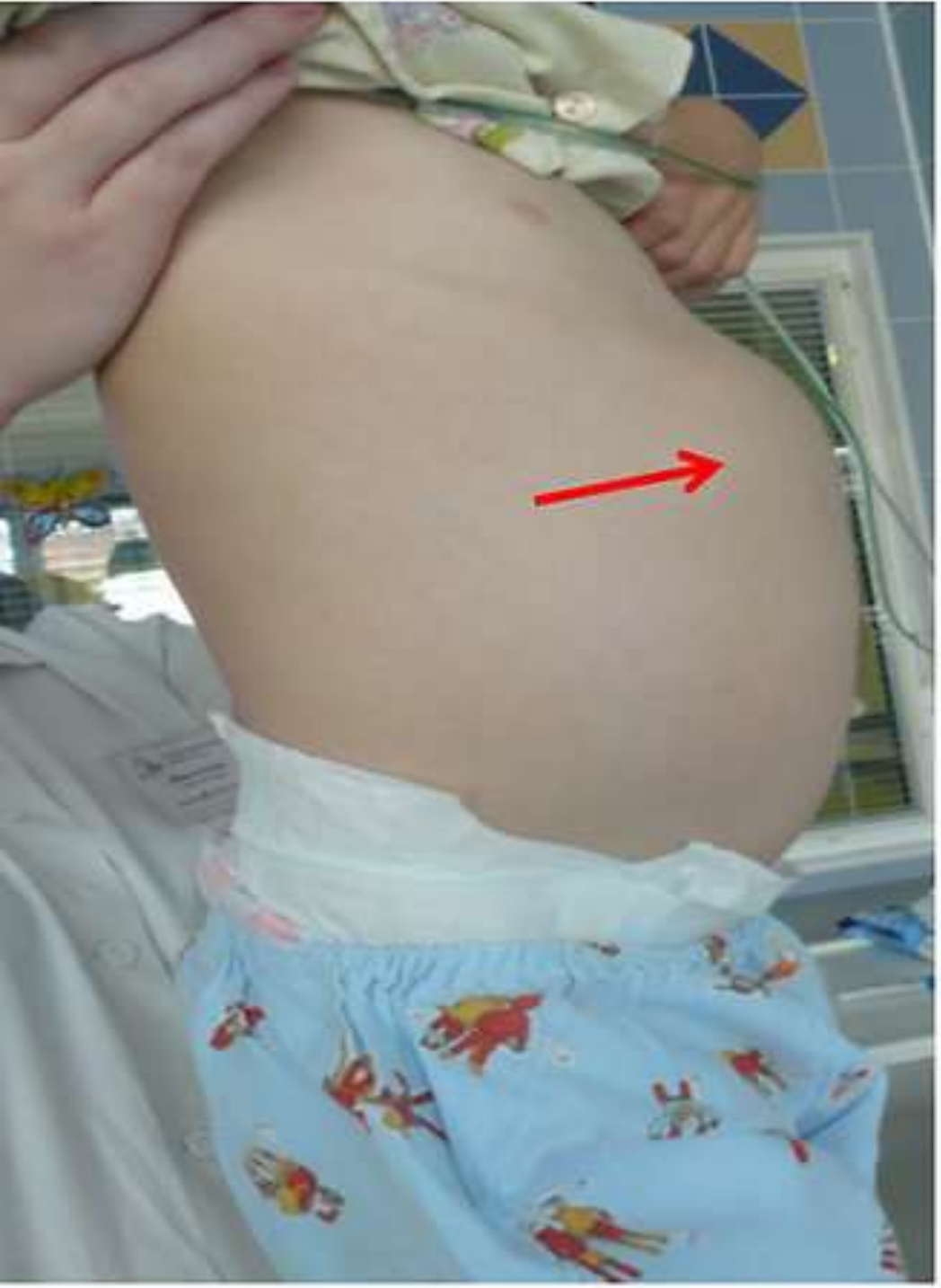
Genetics

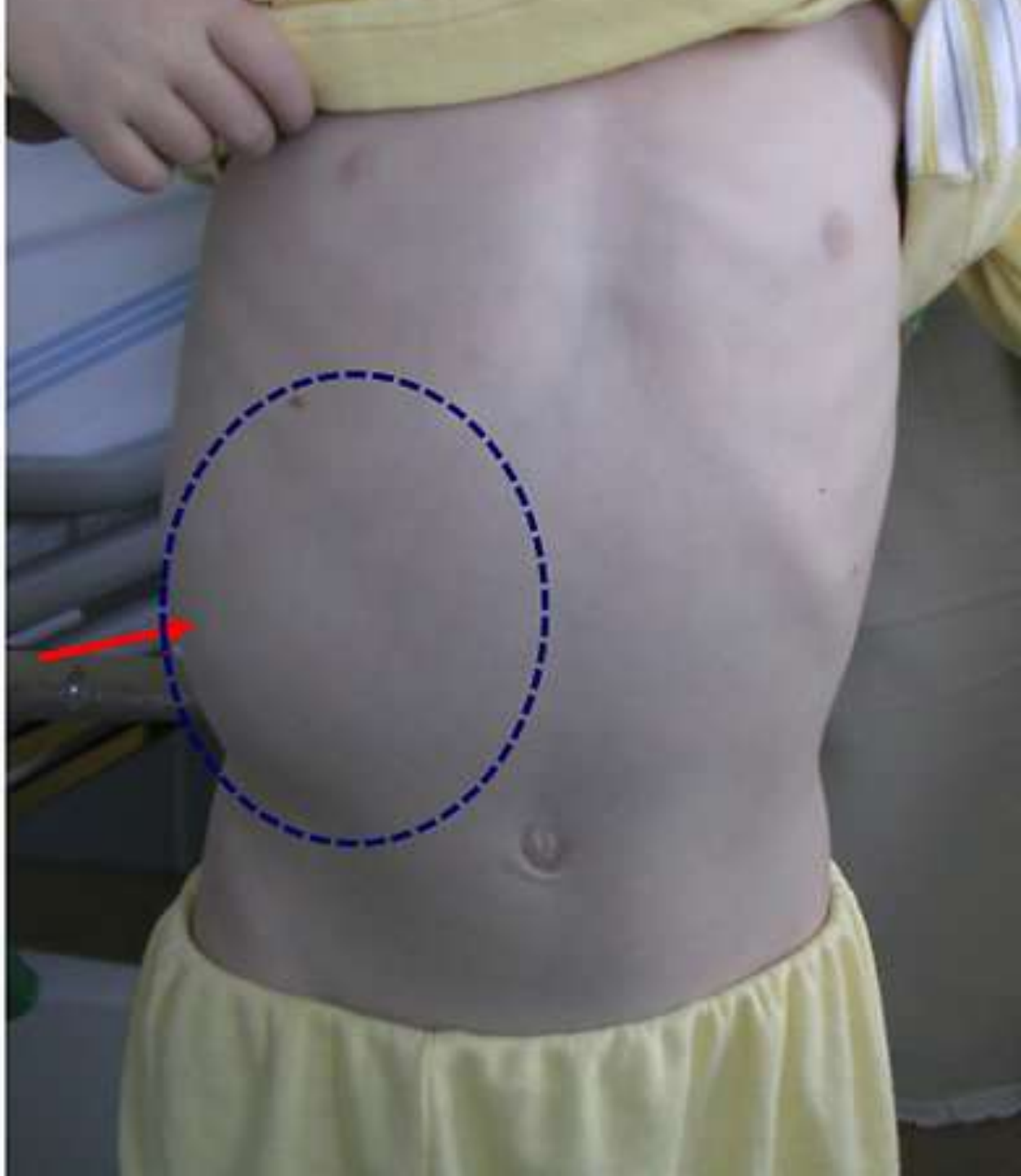
- *WT1* (11p13)
  - Denys-Drash syndrome
  - WAGR syndrome
- *TP53* (17p13)
- *FWT1* (17q12)
- *WT2* (11p15)
  - Beckwith-Wiedemann syndrome
- Loss of heterozygosity at 16q, 1p
- *FWT2* (19q13.3)

F. Netter M.D.

Common symptoms







INTRODUCTION:

- WILMS TUMOR ,NEPHERONMA OR NEPHRON BLASTOMA IS THE MOST FREQUENT INTRA ABDOMINAL TUMOR OF CHILDHOOD AND THE MOST COMMON TYPE OF RENAL CANCER IT IS NAMED FOR MAX WILMS,A GERMAN SURGEON (1867-1981)AND ALSO KNOWN AS NEPHEROBLASTOMA

DEFININATION:-

- WILMS TUMOR IS A CANCEROUS TUMOR OF THE KIDNEY THAT USUALLY OCCURS IN YOUNG CHILDERN

INCIDENCE:-

ITS FREQUENCEY IS ESTIMATED TO 1 IN 50,000 LIVE BIRTH

CAUSES :-

- THE EXACT CAUSE IS NOT KNOWN WILMS TUMOUR PROBABLY ARISES FROM AMALIGNANT, UNDIFFERENTIATED CLUSTER OF PRIMITIVE CELLS CAPABLE OF INITIATING THE REGENERATION OF AN ABNORMAL STRUCTUTLERIDITY

WT gene

- *WT1 was originally considered to be a classic tumor suppressor gene*, and the loss of both copies or mutations of this gene would lead to Wilms tumor development (Rauscher, 1993). Although this may be the case for some tumors, only 20% of patients with Wilms tumor have a mutation in the germline or in tumor tissue.
- *WT2 gene has been linked to the **BWS** ;* excess growth at the cellular, organ (macroglossia, nephromegaly, hepatomegaly), or body segment (hemihypertrophy)
- WTX was found to be inactivated in up to one third of Wilms tumors (Rivera et al, 2007).

PATHOPHYSIOLOGY:-

WHEN AN UNBORN BABY IS DEVELOPING, THE KIDNEYS ARE FORMED FROM PRIMITIVE CELLS

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THE CELLS MATURE &ORGANIZE INTO THE NORMAL KIDNEY STRUCTURE

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SOMETIMES ,CLUMPS OF THESE CELLS REMAIN IN THEIR ORIGINAL, PRIMITIVE FORM

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IF THESE CELLS BEGIN TO MUITIPLY AFTER BIRTH

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THEY MAY UITIMATELY FORM A LARGE MASS OF ABNORMAL CELLS.

↓

THIS IS KNOWN AS A WILMS TUMOR.

CLINICAL MANIFISTATION

- THE USUAL PRESENTATION IS AN ASYMPTOMATIC ABDOMINAL MASS DECTECTED BY THE PARENTS WHILE BATHING THE CHILD
- ABDOMINAL PAIN
- HEMATURIA AS TUMOR EXTENSION IN THE RENAL PELVIS
- HYPERTENSION OCCURS OCCACIONALLY OF EXCESS AMT OF RENIN BY
- TUMOUR
- FEVER
- ANAEMIA
- WEIGHT LOSS
- ANOREXIA
- VOMITING

- ON EXAMINATION, MASS WITHIN THE ABDOMEN THE MASS IS CHARACTERIOLICALLY FIRM , NON-TENDER CONFIRMED TO THE MIDLINE

- IF METASTASIS HAS OCCURRED { THE TUMOR EXTENDS THOUGH THE KIDNEY CAPSULE OR RENAL VEIN &THEN SPREADS TO THE OTHER AREAS OF THE BOBY THROUGH CIRCULATORY SYSTEM}.
  SYMPTOMS OF LUNG INVOLVEMENT SUCH AS DYSPNEA, COUGH, SHORTNESS OF BREATH & PAIN IN THE CHEST MAY BE PRESENT.

DIAGNOSIS:-

- HISTORY

- PHYSICAL EXAMINATION:- CHILDREN WITH WILMS TUMOR GENERALLY FIRST PRESENT TO PHYSICIANS WITH A SWOLLEN ABDOMEN OR WITH AN OBVIOUS ABDOMINAL MASS .

- BLOOD ANALYSIS:- IN THE FORM OF
  - -Hb%
  - -WBC
  - -PLATELET COUNT
  - -LFT
  - -RFT

- URINALYSIS INITIAL DIAGNOSIS OF THE WILMS TUMOR IS MADE BY LOOKING AT THE TUMOR USING VARIOUS IMAGING TECHNIQUES.

- ULTRASOUND ABDOMEN

- C T SCAN { COMPUTED TOMOGRAPHY SCAN}

- INTRAVENOUS PYELOGRAPHY, WHERE A DYE INJECTED INTO A VEIN HELPS SHOW THE STRUCTURES OF THE KIDNEY.

- BIOPSY FINAL DIAGNOSIS HOWEVE R ,DEPENDS ON OBTAINING A TISSUE SAMPLE FROM THE MASS (BIOPSY) THIS BIOPSY IS USUALLY DONE DURING SURGERY TO REMOVE OR DECREASE THE SIZE OF THE TUMOR

- OTHER STUDIES CHEST –XRAY ,CT SCAN OF THE LUNGS,BONE MARROW BIOPSY MAY
  ALSO BE DONE IN ORDER TO SEE IF THE TUMOR HAS SPREAD TO OTHER LOCATION.

- MRI

STAGING

- STAGE 1: - TUMOR CONFINED TO KID NEY & COMPLETELY EXCISED .

- STAGE 2 : - TUMOR EXTENDS BEYOND KIDNEY BUT COMPLETELY EXCISED

- STAGE 3: - TUMOR INFILTRATES RENAL FAT RESIDUAL TUMOR AFTER SURGERY LYMPHNODE INVOLVEMENT OF HILUM ,PARA-AORTIC OR BEYOND

- STAGE 4:- METASTASIS IN LUNG OR LIVER RARELY IN BONE AND BRAIN

- STAGE 5:- BILATERAL RENAL INVOLVEMENT

- **HISTOLOGY:-** ARE THE TYPES OF CANCER CELL WITHIN THE WILMS TUMOR

- **FAVOURABLE:-** IT MEANS THEY RESPOND WELL TO CONVENTIONAL THERAPY

- **UNFAVOURABLE:-** ITS MEANS THEY ARE MORE AGGRESSIVE CANCER TYPES AND DO NOT RESPOND WELL TO CONVENTIONAL THERAPY.

❑1. SURGERY:-

AN OPERATION TO REMOVE THE CANCER

(A) APARTIAL NEPHRECTOMY:- IS WHEN THE CANCER AND PART OF THE KIDNEY ARE REMOVED .ITS USUAIIL DONE IF THE OHER KIDNEY IS DAMAGED OR HAS ALREADY BEEN REMOVED

(B) A SIMPLE NEPHRECTOMY :-IS WHEN THE WHOLE KIDNEY IS REMOVED

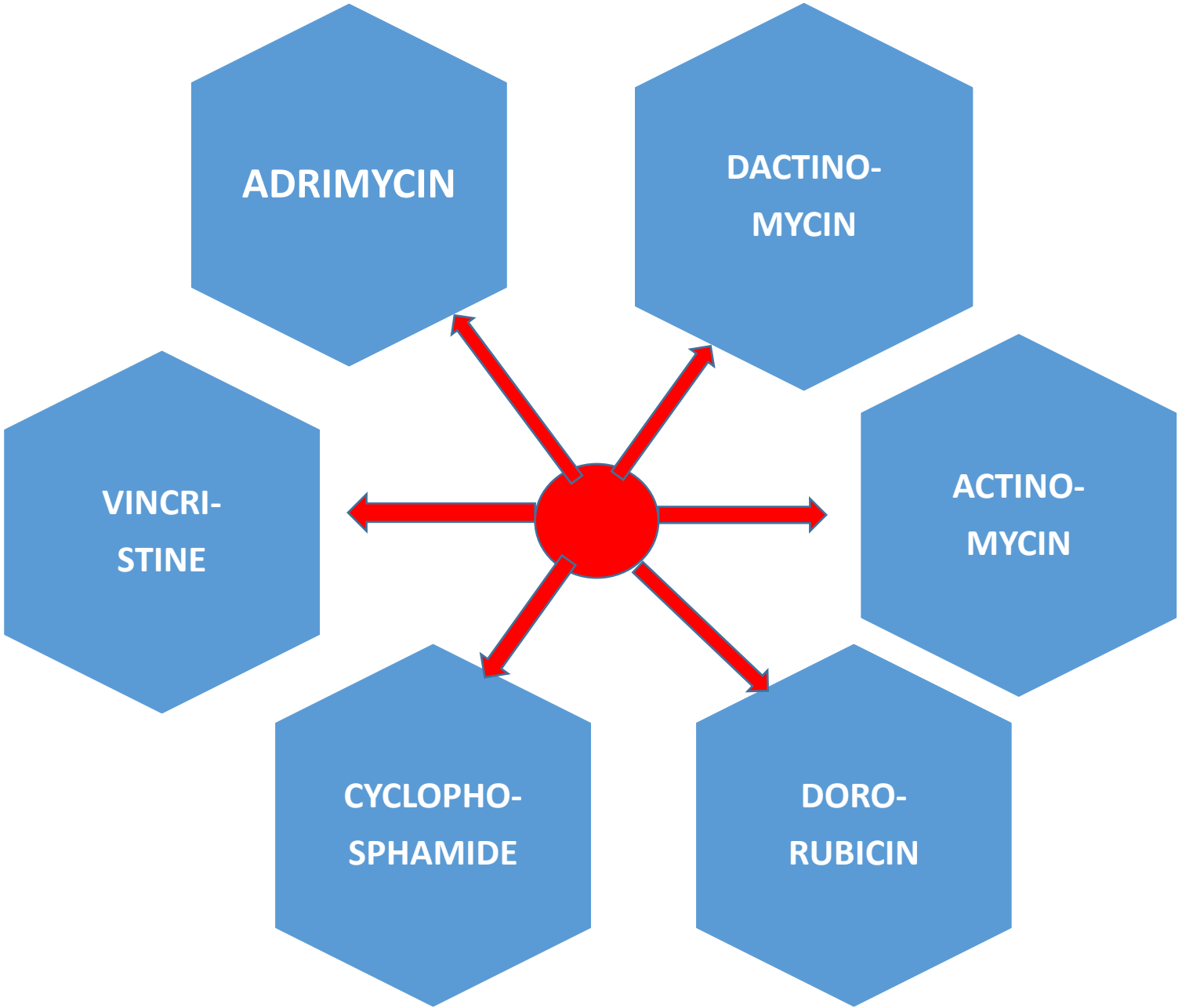
(C) A RADICAL NEPHRECTOMY :-REMOVES THE ENTIRE KIDNEY AND TISSUE AROUND IT. SOME TIME SOME LYMPH NODES MAY ALSO BE REMOVED.

❑2.CHEMOTHERAPY:-

- ❑MEDICATION USED TO KILL CANCER CELLS .IT IS ALSO CALLED AS SYSTEMIC TREATMENT BECAUSE THE DRUGS ENTER THE BLOOD AND KILL CANCER CELL THROUGHOUT THE BODY.

- ❑CHEMOTHERAPY GIVEN AFTER AN OPERATION WHEN THERE ARE NO CANCER CELL THAT CAN BE CALLED AS ADJUVANT THERAPY.

DRUGS


ADRIMYCIN
DACTINO-
MYCIN
VINCRI-
STINE
ACTINO-
MYCIN
CYCLOPHO-
SPHAMIDE
DORO-
RUBICIN

❑(3) RADIATION THERAPY:-

- ❑X-RAYS OR OTHER HIGH ENREGY RAYS ARE USED TO KILL CANCER CELLS AND SHRINK TUMORS.IT MAY BE USED BEFORE AND AFTER SURGERY AND /OR CHEMOTHERAPY.

RADIATION THERAPY IS NOT GIVEN IN CHILDREN BELOW AGE OF 1YRS.

❑STANDARD TREATMENT OF WILMS TUMOR

❑TREAMENT DEPENDS ON THE STAGE OF THE DISEASE,

❑THE HISTOLOGY OF THE CELL TYPE AND THE PATIENT AGE AND THE GENERAL HEALTH IN EACH CASE IS UNIQUE AND THERE MAY BE SITUATION WHERE IT IS NECESSARY OR DESIRABLE TO DEVIATE FROM THE STANDARD COURSE OF TREATMENT.

STAGE 1:-

IF YOUR CHILD HAS A FAVOURABLE HISTOLOGY TUMOR , YOUR CHILD TREATMENT WILL PROBLY BE SURGERY TO REMOVE THE CANCER FOLLWED BY CHEMOTHERAPY .

IF YOUR CHILD HAS AN UNFAVOURABLE TUMOR YOUR CHILDS TREATMENT WILL PROBABLY BE SURGERY TO REMOVE THE CANCER FOLLWED BY RADIATION THERAPY PLUS CHEMOTHERAPY

- **STAGE 2:-**

- IF YOUR CHILD HAS A FAVOURABLE HISTOLOGY TUMOR ,

- YOUR CHILD TREATMENT WILL PROBABLY BE SURGERY TO REMOVE THE CANCER FOLLWED BY CHEMOTHERAPY

- IF YOUR CHILD HAS AN UNFAVOURABLE HISTOLOGY TUMOR ,

- YOUR CHILDS TERATMENT WILL PROBABLY BE SURGERY TO REMOVE THE CANCER FOLLWED BY RADIATION THERAPY PLUS CHEMOTHERAPY.

STAGE 3:-

- IF YOUR CHILD HAS A FAVOURABLE OR AN UNFAVOURABLE HISTOLOGY TUMOR ,

- YOUR TREATMENT WILL PROBABLE BE SURGERY TO REMOVE THE CANCER FOLLWED BY RADIATION THERAPY PULS CHEMOTHERAPY.

- SOMETIMES THE CANCER CANNOT BE REMOVED DURING SURGERY BECAUSE IT IS TOO CLOSE TO IMPORTANT ORGANS OR BLOOD VESSELS OR BECAUSE IT IS TOO LARGE TO REMOVE

- IN SUCH CASE THE DOCTOR MAY ONLY PERFORMED A BIOPSY AND THEN GIVEN CHEMOTHERAPY WITH OR WITHOUT RADIATION THERAPY ONCE THE CANCER HAS BECAME SAMLLER ,

- SURGERY CAN BE PERFORMED ,FOLLOWED BY ADDITIONAL

- RADIATION THERAPY AND CHEMOTHERAPY

STAGE 4:-

- IF YOUR CHILD HAS A FAVOURABLE OR UNFAVOURABLE
- HISTOLOGY TUMOR YOUR CHILDS TREATMENT WILL PROBABLY BE SURGERY TO REMOVE THE CANCER FOLLOWED BY RADIATION THERAPY.
- PT’S WHOSE CANCER HAS METASTASIS TO THE LUNGS WILL RECIVE ADDITIONAL CHEMOTHERAPY .

STAGE 5:-

- BECAUSE BOTH KIDNEYS CONTAIN CANCER IT IS NOT USUALLY POSSIBLE TO REMOVE BOTH KIDNEYS YOUR CHILDS DOCTOR WILL PROBABLY TAKE OUT A PIECE OF THE CANCER IN BOTH KIDNEYS AND REMOVE SOME LYMPH NODES AROUND THE KIDNEY TO SEE WHETHER THEY CONTAIN CANCER.

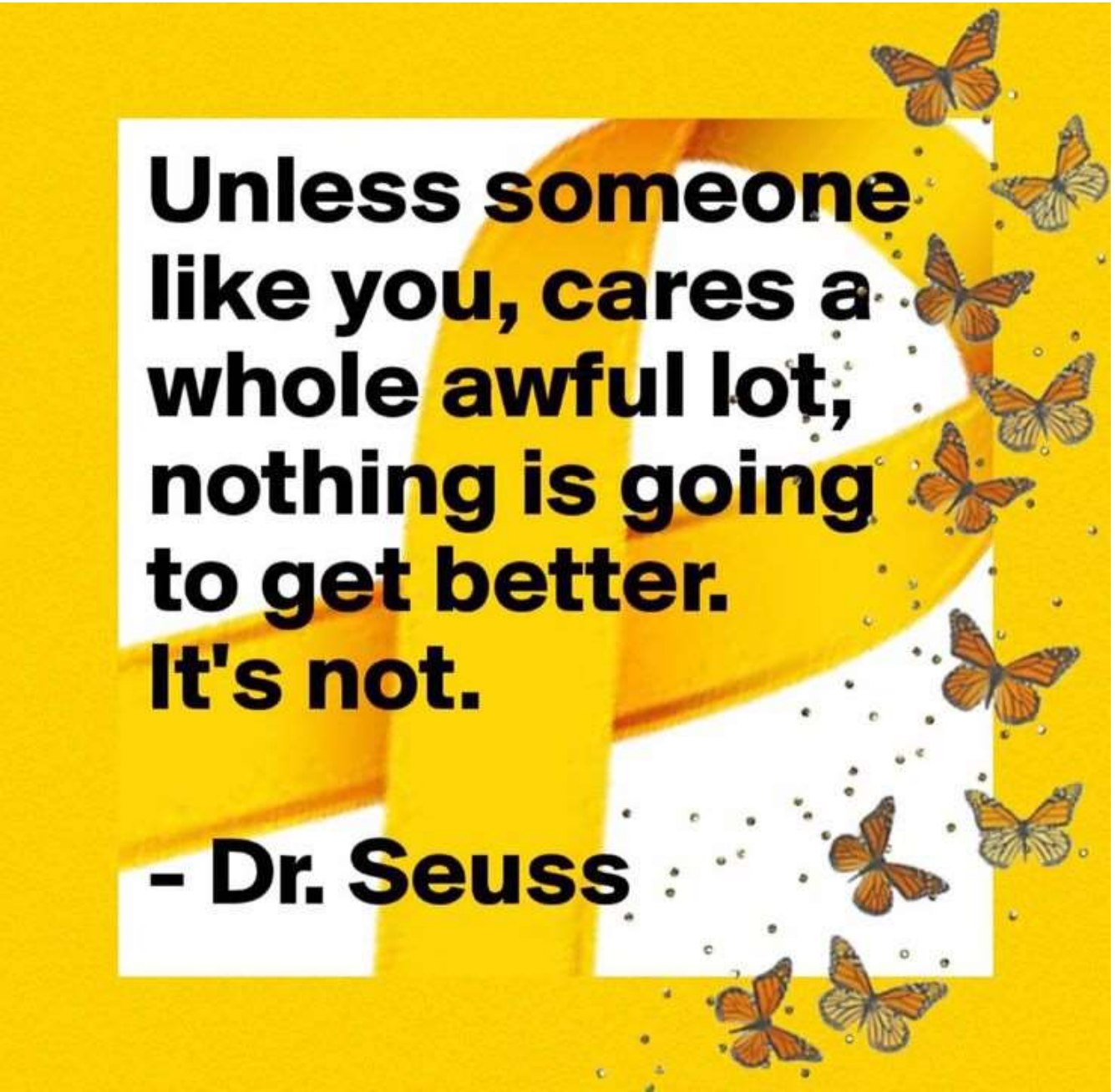
- FOLLOWING SURGERY, CHEMOTHERAPY IS GIVEN TO SHRINK THE CANCER .

- A SECOND OPERATION IS THEN PERFORMED TO REMOVE AS MUCH OF THE CANCER AS POSSIBLE WHILE LEAVING AS MUCH OF THE KIDNEYS AS POSSIBLE.SURGERY MAY BE FOLLOWED BY MORE CHEMOTHERAPY AND /OR RADIATION THERAPY

PROGNOSIS:-

- IS THE PREDICTION OF THE COURSE AND OUT COME OF THE DISEASE. FOR WILMS, THE PROGNOSIS IS GENERALLY VERY GOOD.MORE THAN 85%CURE RATE. QUITE A BIT DEPENDS, THOUGH ON THE STAGING OF THE TUMOUR AND THE HISTOLOGY OF THE TUMORE

THANK YOU


Unless someone like you, cares a whole awful lot, nothing is going to get better. It's not.
- Dr. Seuss