مؤسسة مستشفى سرطان الأطفال-مصر
Children's Cancer Hospital Foundation - Egypt
57357

Adrenocortical carcinoma disease

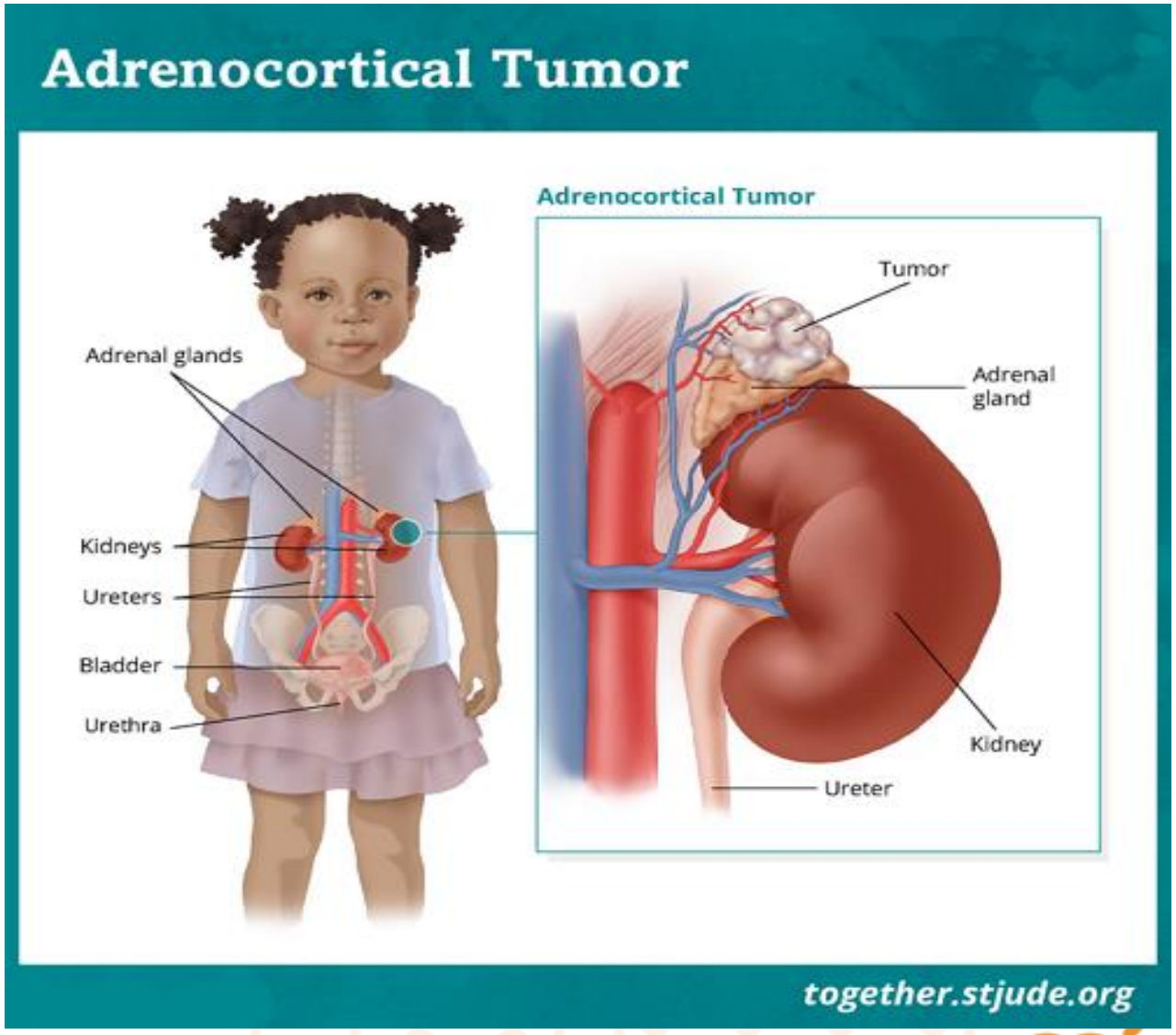
ACT

BY: Bodour Ahmed

- Adrenocortical tumors (ACT) constitute a rare but aggressive malignancy in children.

- A wide spectrum of germline TP53 alterations have been described in ACT.

- Cancer of the adrenal glands, which are two small triangular-shaped glands that sit on top of each kidney. **The outer layer** of the adrenal gland is called the adrenal cortex. **The adrenal cortex** produces male and female sex hormones called **androgens** and **estrogens**. These hormones affect the development of male and female traits.



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Hormone-related signs and symptoms of pediatric Adrenocortical Tumor (ACT)

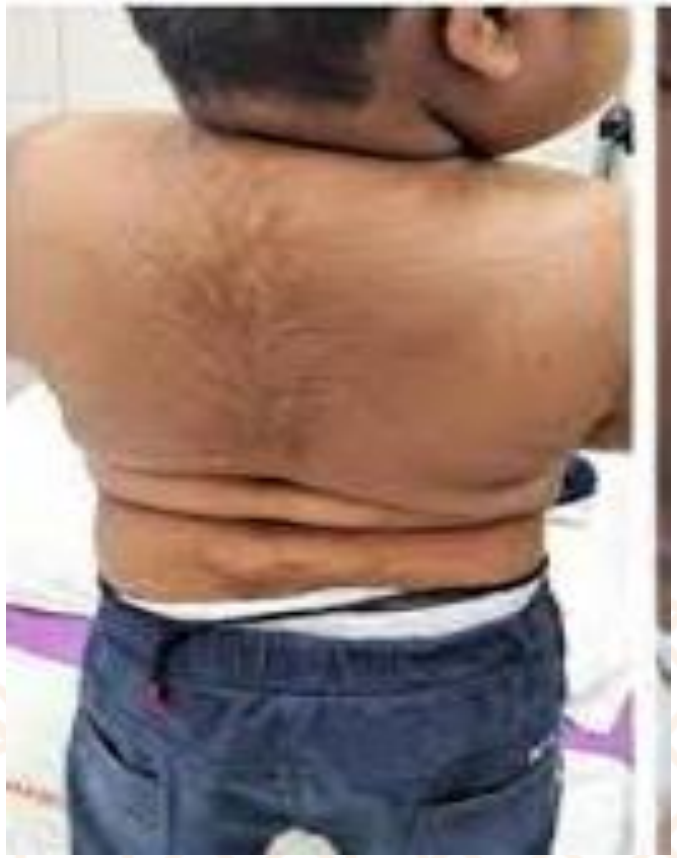
Androgen	Estrogen	Cortisol	Aldosterone
Early puberty, male traits such as facial and body hair, acne, deepening voice, increased growth.	Early puberty, female traits such as breast growth.	Rounded face, weight gain, fatty hump on upper back, stunted height, high blood sugar, high blood pressure.	High blood pressure, thirst, muscle cramps.

❑ Cushing syndrome (↑Cortisol)


Fig.1a

Fig1b

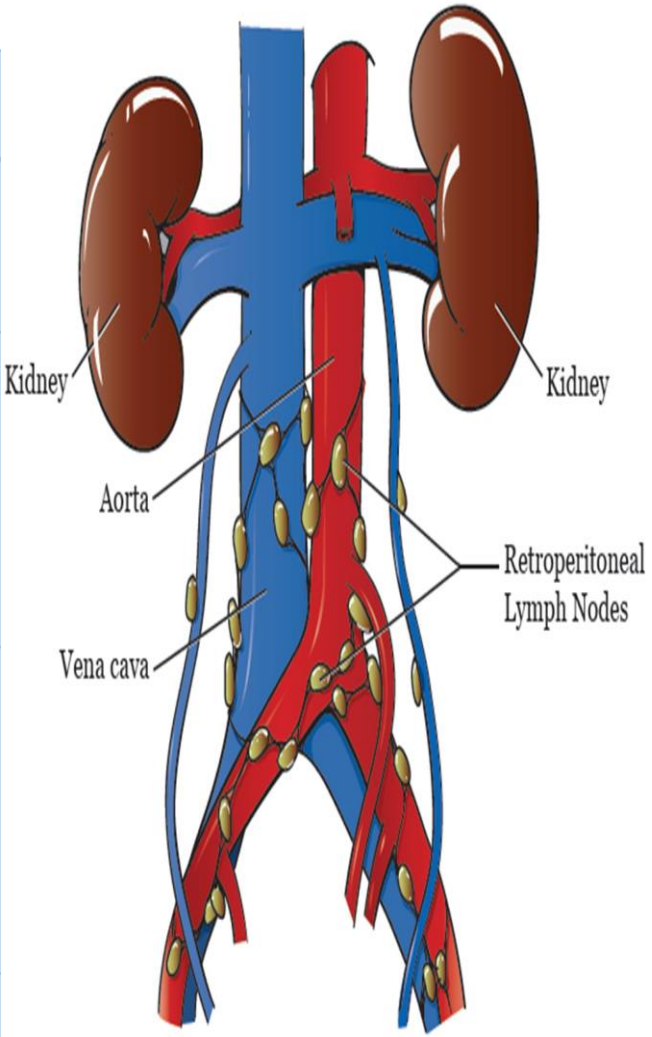
❑ Virilization (↑ Androgens)



❑ Diagnosis of ACT:

- ➢ **Laboratory test (blood and urine) for hormonal levels.**
- ➢ **CT & MRI.**
- ➢ **PET scan.**

Stage	symptoms
I	Completely resectable , small tumors (< 200 cm3 and<100 g) with normal postoperative hormone levels for 1month.
II	Completely resectable, large tumors (≥ 200 cm3 and ≥100g) with normal postoperative hormone levels& will undergo extended regional lymph node dissection.
III	➢ Unresectable, gross or microscopic residual disease tumor spillage. ➢ Patients with stage I & stage II of tumors fail to normalize hormone levels after surgery.
IV	Presence of metastatic disease.



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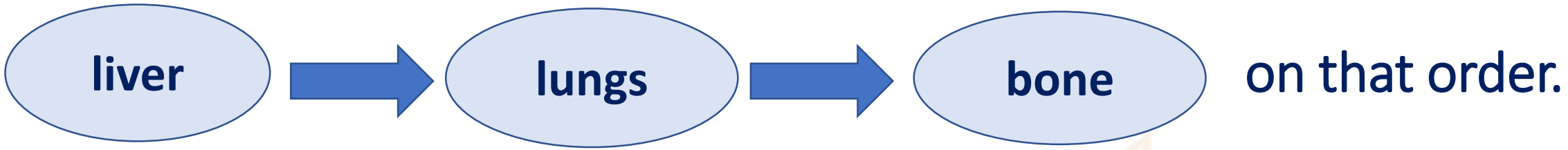
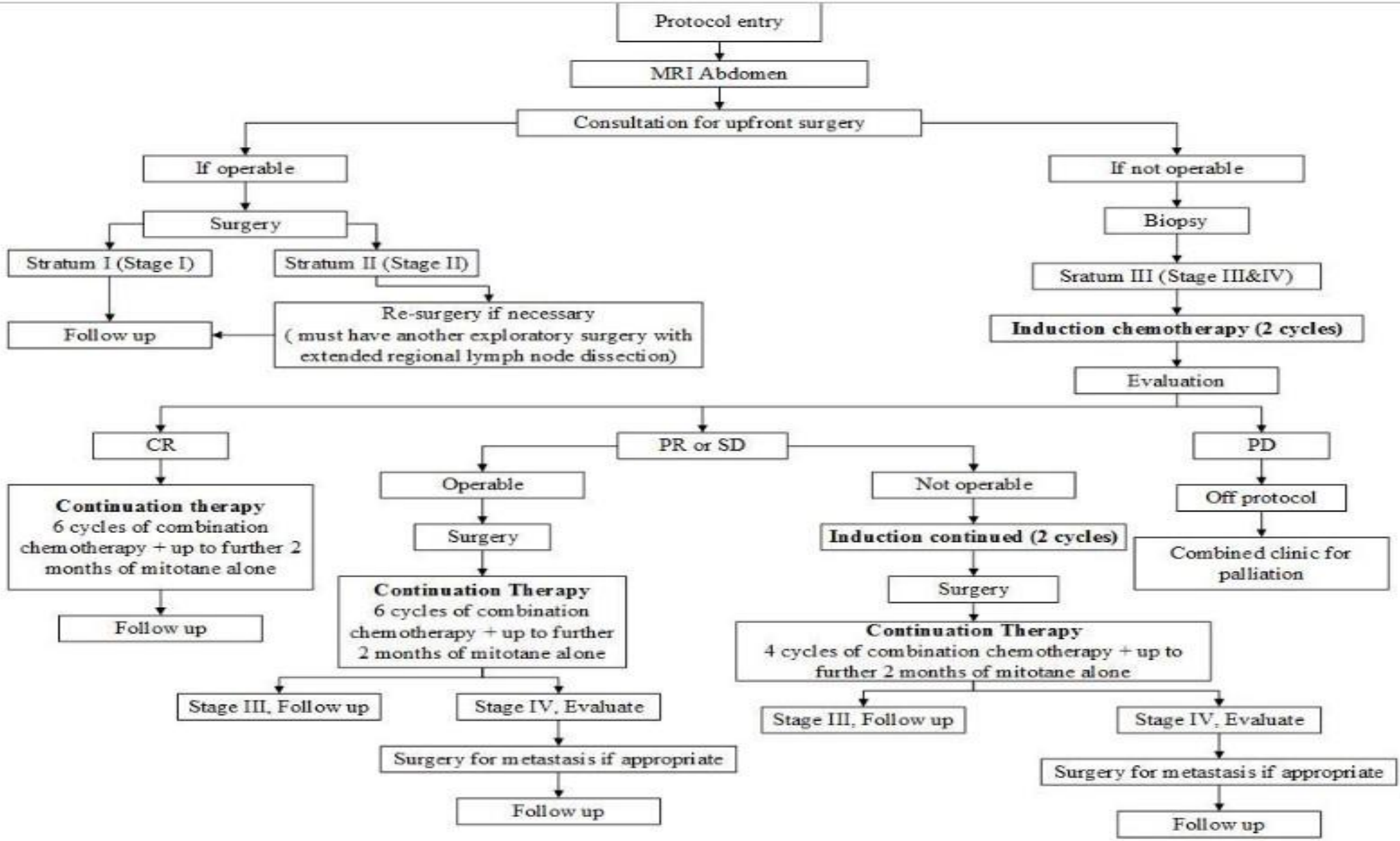
➢Metastasis:



on that order.



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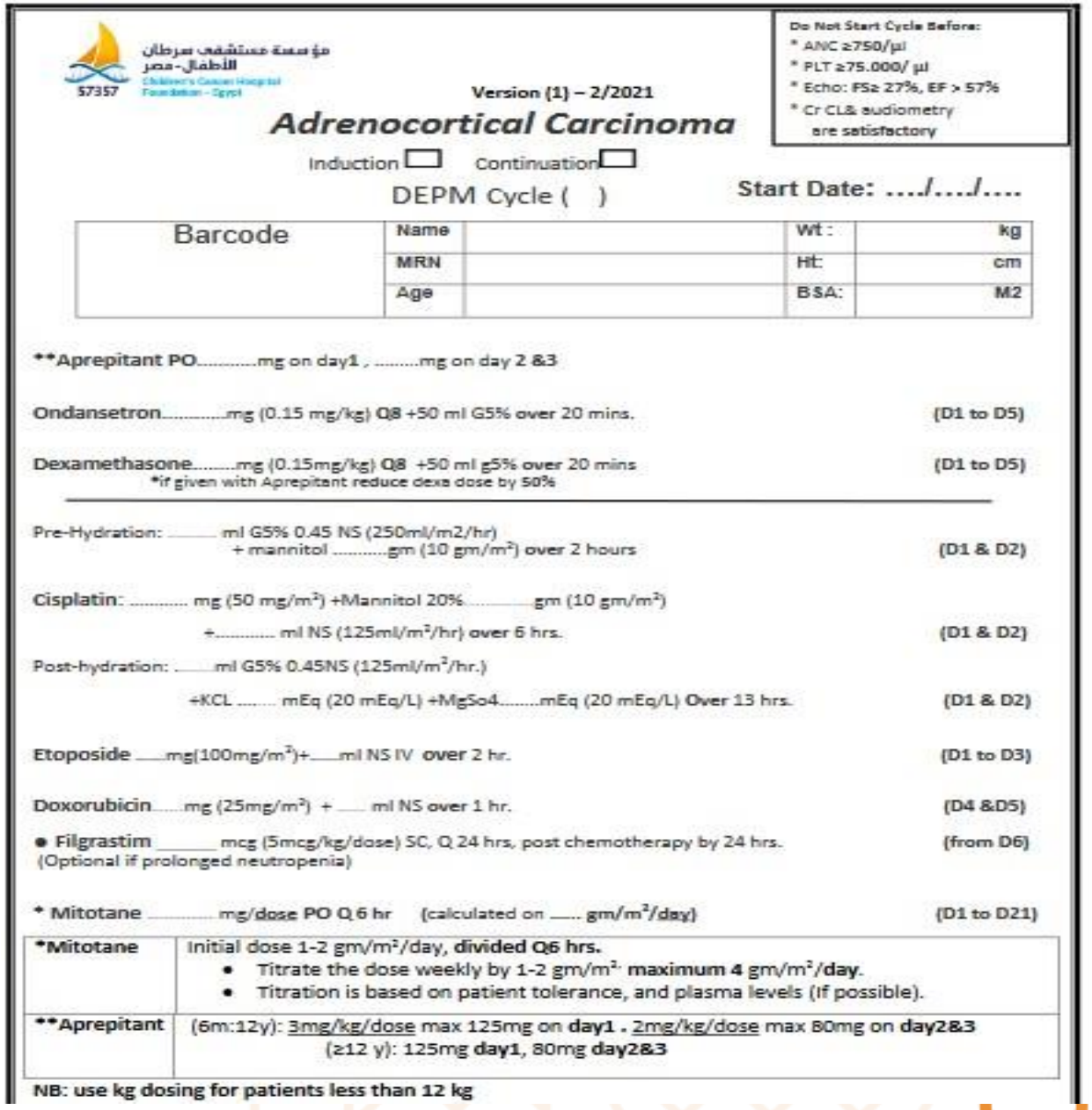
- **Total Number of chemotherapy cycles is maximum 8 cycles (2-4 Induction + 4-6 Continuation). Patient will continue with mitotane alone for up to a further 8 weeks & each cycle consists of 21 days.**

  - **Evaluation: after cycle 2, 4, 6, 8.**
  - **Surgery: after cycle 2 or 4.**



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Version (1) – 2/2021

Adrenocortical Carcinoma

Do Not Start Cycle Before:
* ANC ≥750/µl
* PLT ≥75.000/ µl
* Echo: FS≥ 27%, EF > 57%
* Cr CL& audiometry are satisfactory

Induction ☐ Continuation ☐

DEPM Cycle ( )

Start Date: …./…./….

Barcode	Name		Wt :	kg
	MRN		Ht:	cm
	Age		BSA:	M2

**Aprepitant PO……mg on day1 , ……mg on day 2 &3

Ondansetron……mg (0.15 mg/kg) Q8 +50 ml G5% over 20 mins. (D1 to D5)

Dexamethasone……mg (0.15mg/kg) Q8 +50 ml g5% over 20 mins (D1 to D5)
*if given with Aprepitant reduce dexa dose by 50%

Pre-Hydration: …… ml G5% 0.45 NS (250ml/m2/hr)
+ mannitol ……gm (10 gm/m²) over 2 hours (D1 & D2)

Cisplatin: …… mg (50 mg/m²) +Mannitol 20%……gm (10 gm/m²)
+…… ml NS (125ml/m²/hr) over 6 hrs. (D1 & D2)

Post-hydration: ……ml G5% 0.45NS (125ml/m²/hr.)
+KCL …… mEq (20 mEq/L) +MgSo4……mEq (20 mEq/L) Over 13 hrs. (D1 & D2)

Etoposide ……mg(100mg/m²)+……ml NS IV over 2 hr. (D1 to D3)

Doxorubicin……mg (25mg/m²) + …… ml NS over 1 hr. (D4 &D5)

● Filgrastim _____ mcg (5mcg/kg/dose) SC, Q 24 hrs, post chemotherapy by 24 hrs. (from D6)
(Optional if prolonged neutropenia)

* Mitotane …… mg/dose PO Q 6 hr (calculated on …… gm/m²/day) (D1 to D21)

*Mitotane	Initial dose 1-2 gm/m²/day, **divided Q6 hrs.** • Titrate the dose weekly by 1-2 gm/m². **maximum 4** gm/m²/**day**. • Titration is based on patient tolerance, and plasma levels (If possible).
**Aprepitant	(6m:12y): 3mg/kg/dose max 125mg on **day1** . 2mg/kg/dose max 80mg on **day2&3** (≥12 y): 125mg **day1**, 80mg **day2&3**

NB: use kg dosing for patients less than 12 kg

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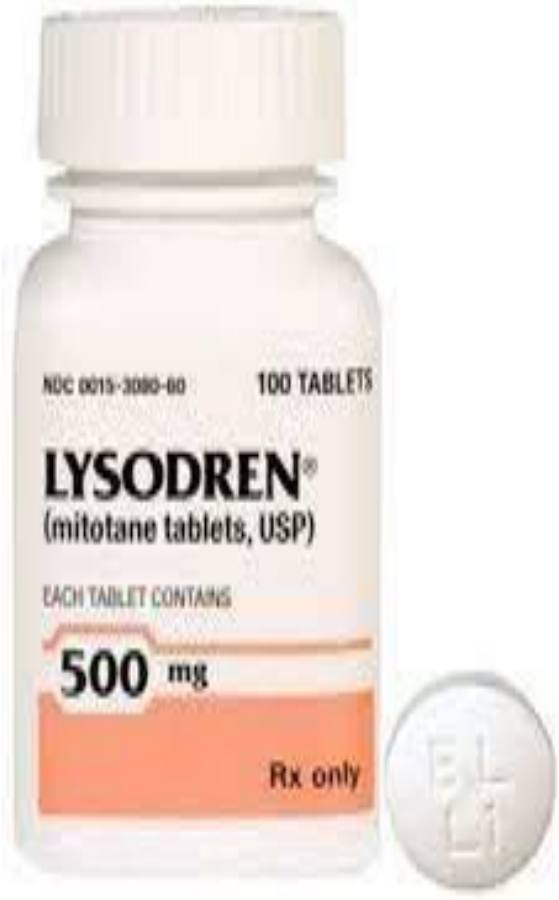
Mitotane



- **Mitotane** is antineoplastic agent (directly) destroys the mitochondria resulting in necrosis of the adrenal cortex. &suppresses the secretion of adrenal steroids.
- Goal steady-state plasma levels are 14 and 20micrograms/mL. Therapeutic levels are achieved after approximately 14 weeks of therapy.
- If interruption of mitotane for 4-5 days occurs for toxicity, for example, severe gastrointestinal toxicity, it is recommended to reduce the dose of mitotane to 2/3rd of the previous dose.

References;

- https://research.57357.org/protocols/?dir=uploads/protocols/57357Protocols/Adreno-cortical-Carcinoma.
- https://online.lexi.com/lco/action/doc/retrieve/docid.

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AFNCI

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