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ADRENOCORTICAL TUMORS – AN AUDIT FROM A TERTIARY CARE CENTRE

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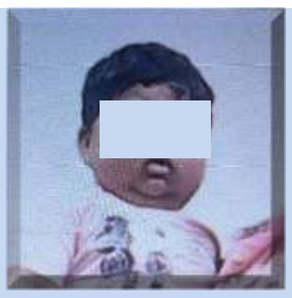
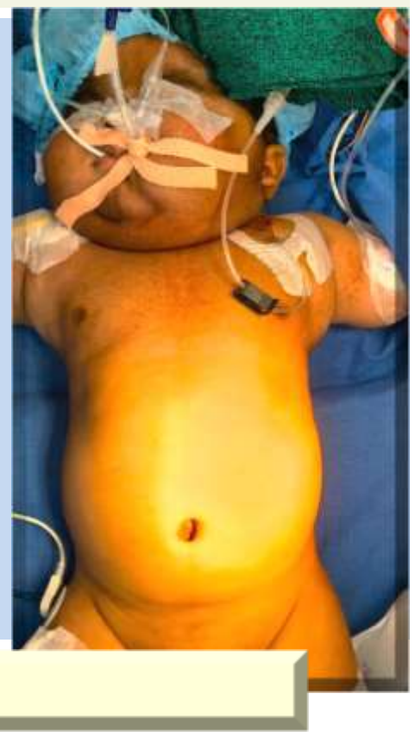
INTRODUCTION

- **Rare, accounting for <0.2% of pediatric malignancies.[1]**
- **Aggressive, with variable hormonal activity and presentation.[2]**
- **Linkage to hereditary cancer syndromes like Li-Fraumeni syndrome.[3]**
- **Complete surgical resection (R0) provides the best chance of cure.**
  Challenges - tumor size, vascular invasion and local extension.
- Early diagnosis and multidisciplinary management are key for improved outcomes.





AIMS: **To review our experience in the management and outcome of ACC at our institute.**

Methodology

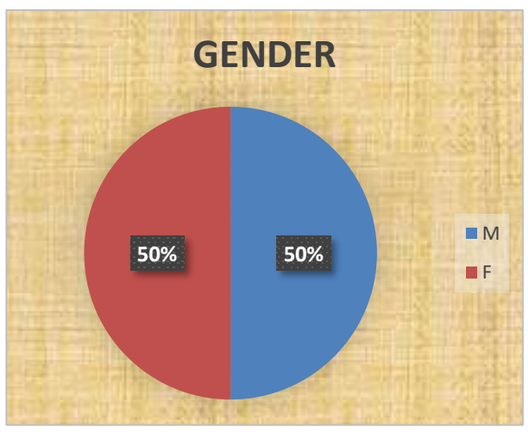
Study Design - Retrospective

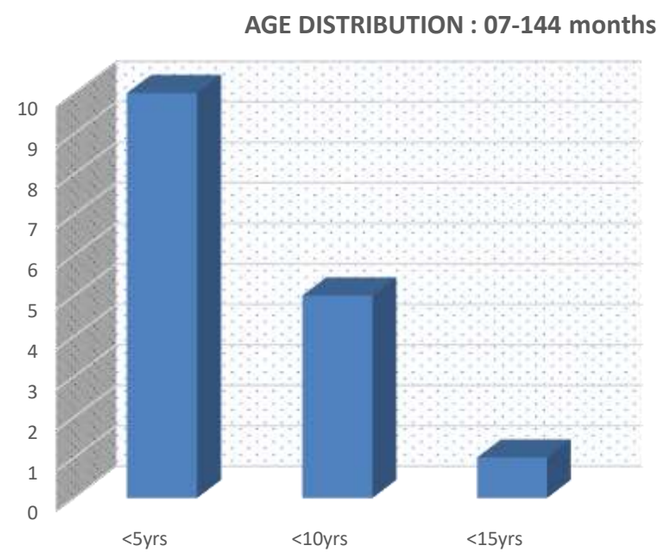
Duration – Jan 2012 – Jan 2025

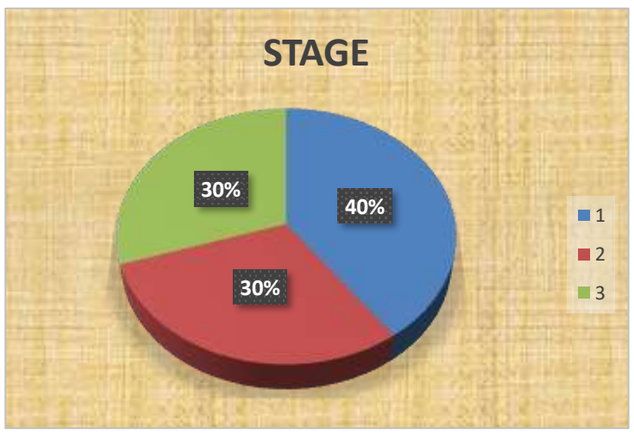
Inclusion Criteria- ≤15 yrs operated for ACC-TMH

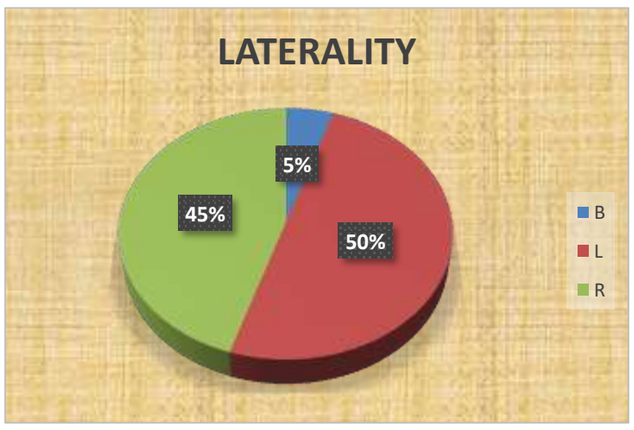
Exclusion Criteria
- Non-operated or outside-operated patients.
- Secondary adrenal involvement
- Non Consent to Surgery.
- Missing records

Results : Total Pt. (n) – 20.









DEC Regimen	No of patient (n= 20)
Adjuvant chemo	12
Observation	5
Abandoned chemo	3

SURGERY TYPE	No (n- 20)
BILATERAL ADRENALECTOMY	01
LEFT ADRENALECTOMY	10
RIGHT ADRENALECTOMY	09
RIGHT NEPHRECTOMY	01
LEFT NEPHRECTOMY	02
IVC THROMBECTOMY	03
LEFT RENAL VEIN THROMBECTOMY	01

Duration of hospital stay : 5 – 19 days [M - 8.5]

Duration of Surgery : 90 – 540 min [M – 187]

Blood Loss : 20 - 4000 ml [Median -100 ml]

COMPLICATIONS

Intra-operative hemorrhage- 2
Renal pedicle injury- 1
Superficial wound infection/dehiscence - 2
Subacute intestinal obstruction - 1
Chyle leak -2
Pancreatitis -1
Atelectasis - 2

FOLLOW UP

Died: 1 Referred: 3

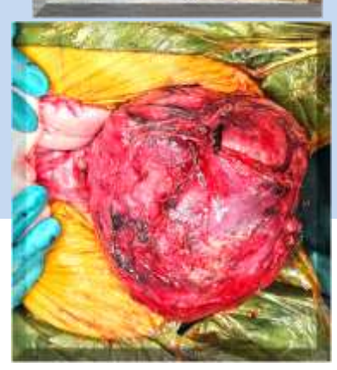
Lost in F/u: 2

Alive: 14

[Without active disease – 9, Recurrences - 5]

TAKE-HOME MESSAGE

- ☐ **Rare but aggressive** – carries significant morbidity, mortality.
- ☐ **Early diagnosis matters** – Prompt recognition of symptoms, hormonal evaluation, imaging studies - vital for timely intervention.
- **Surgery is the cornerstone** – Complete resection (R0) - most important prognostic factor for survival.
- ☐ **Multidisciplinary approach** – Integrated care - Pediatric surgery, Oncology, Endocrinology, Radiology & Pathology optimizes outcome.
- **Long term follow up** – Essential for detecting recurrence & monitoring late effects of therapy.
- **Audit driven improvement** – Institutional reviews helps identify gaps & enhance clinical practice.



References
1 Ribeiro RC, Pinto EM, Zambetti GP, et al. The International Pediatric Adrenocortical Tumor Registry initiative: contributions to clinical, biological, and treatment advances in pediatric adrenocortical tumors. Mol Cell Endocrinol 2012;351(1):37–43.
2 Pandey A et al., Indian J Pediatr Surg. 2019;24(3):182-187
3 Sun-Zhang A, Juhlin C, Carling T, et.al. Comprehensive genomic analysis of adrenocortical carcinoma reveals genetic profiles associated with patient survival. ESMO Open Volume 9, Issue 7, July 2024, 103617.