AMSER Case of the Month October 2024

14-year-old male with bilateral scrotal swelling

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Patient Presentation

- HPI: 14-year-old male presented with bilateral scrotal swelling
- PMH: Patient was diagnosed with CAH at birth on routine newborn screen (17-OHP = 603.00) and genetic testing (homozygous mutation in the 21-OH gene). He was started on prednisone and fludrocortisone, and has been following up regularly in the clinic for dose management. He was diagnosed with precocious puberty in 2012 for which he underwent Histrelin implantation, subsequently removed in 2020.

Patient Presentation

- PSH: Histrelin implant in 2012
- FH: Mother has history of asthma, hypertension, obesity, cholecystectomy, DM and hyperlipidemia. Father has history of DM, hypertension.
- SH: Lives with mother and 5 siblings. Has IEP in school

Patient Presentation cotd.

Physical exam:

- Vitals- BP- 116/68 , Pulse- 80 , Temp- 36.3 °C (97.3 °F)
- Ht- 146.9 cm (4' 9.84")
- Wt- 71.6 kg (157 lb/ 14.4 oz)
- BMI- 33.19 kg/m^2
- Systemic examination: Normal, except for bilateral palpable scrotal masses.

Pertinent Labs

Labs	Reference range	Results
Testosterone Free	0.48 - 15.3 ng/dL	32.2 (H)
Testosterone	100-950 ng/dL	668
Hydroxyprogesterone	9.00 - 140.00 ng/dL	11600.87 (H)
Androstenedione	<1.0 ng/mL	16.7 (H)
Cortisol	4.8 - 19.5 ug/dL	0.8 (L)
LH	1.8 - 10.8 mIU/mL	<0.1 (L)
FSH	1.5 - 12.4 mIU/mL	<0.1 (L)
Inhibin B	47 - 383 pg/mL	80

What Imaging Should We Order?

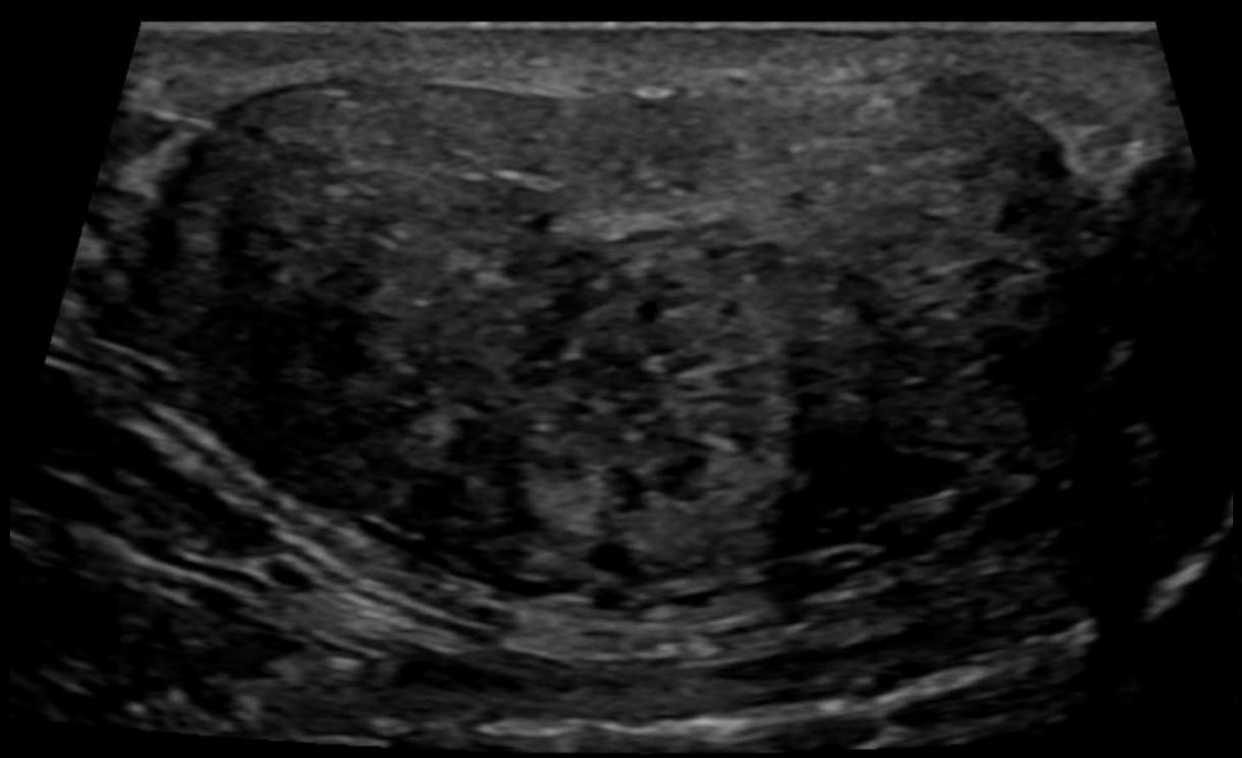
Select the applicable ACR Appropriateness Criteria

Variant 2: **Newly diagnosed palpable scrotal abnormality. No history of trauma or infection. Initial imaging.**

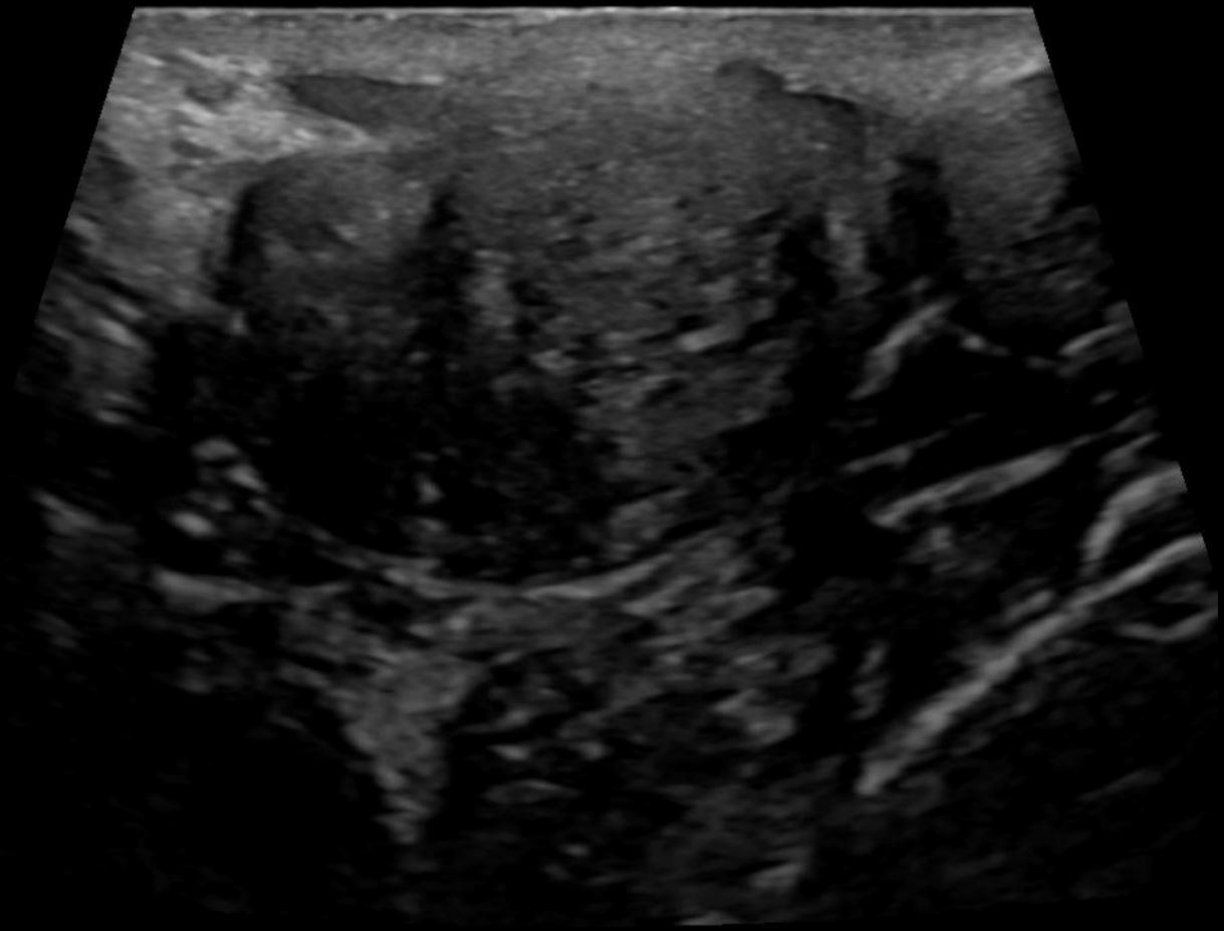
Procedure	Appropriateness Category	Relative Radiation Level
US duplex Doppler scrotum	Usually Appropriate	O
US scrotum	Usually Appropriate	O
MRI pelvis (scrotum) without and with IV contrast	May Be Appropriate	O
MRI pelvis (scrotum) without IV contrast	May Be Appropriate	O
CT abdomen and pelvis with IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	Usually Not Appropriate	☢☢☢
CT pelvis with IV contrast	Usually Not Appropriate	☢☢☢
CT pelvis without IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
MRI abdomen and pelvis without and with IV contrast	Usually Not Appropriate	O
MRI abdomen and pelvis without IV contrast	Usually Not Appropriate	O
Nuclear medicine scan scrotum	Usually Not Appropriate	☢☢☢

This imaging modality was ordered by the physician

Findings (unlabeled)



SAG LT TESTIS LAT-MED



SAG RT TESTIS LAT-MED

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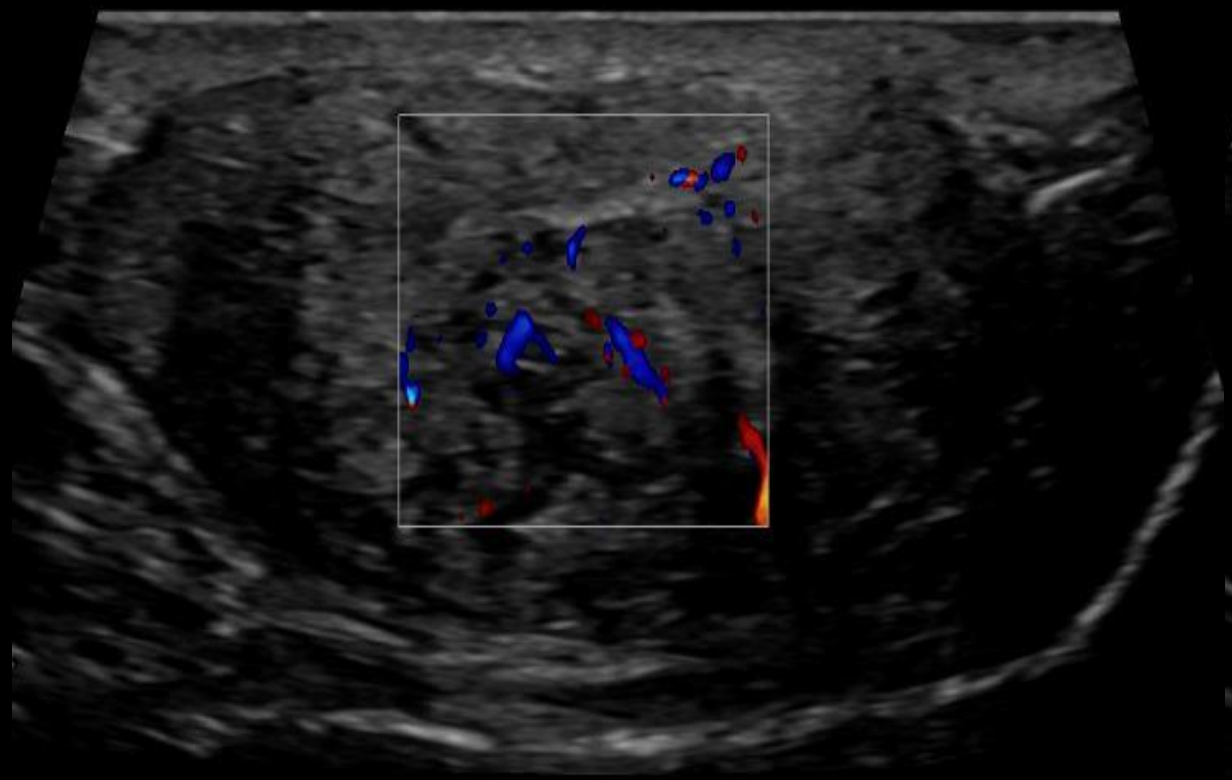
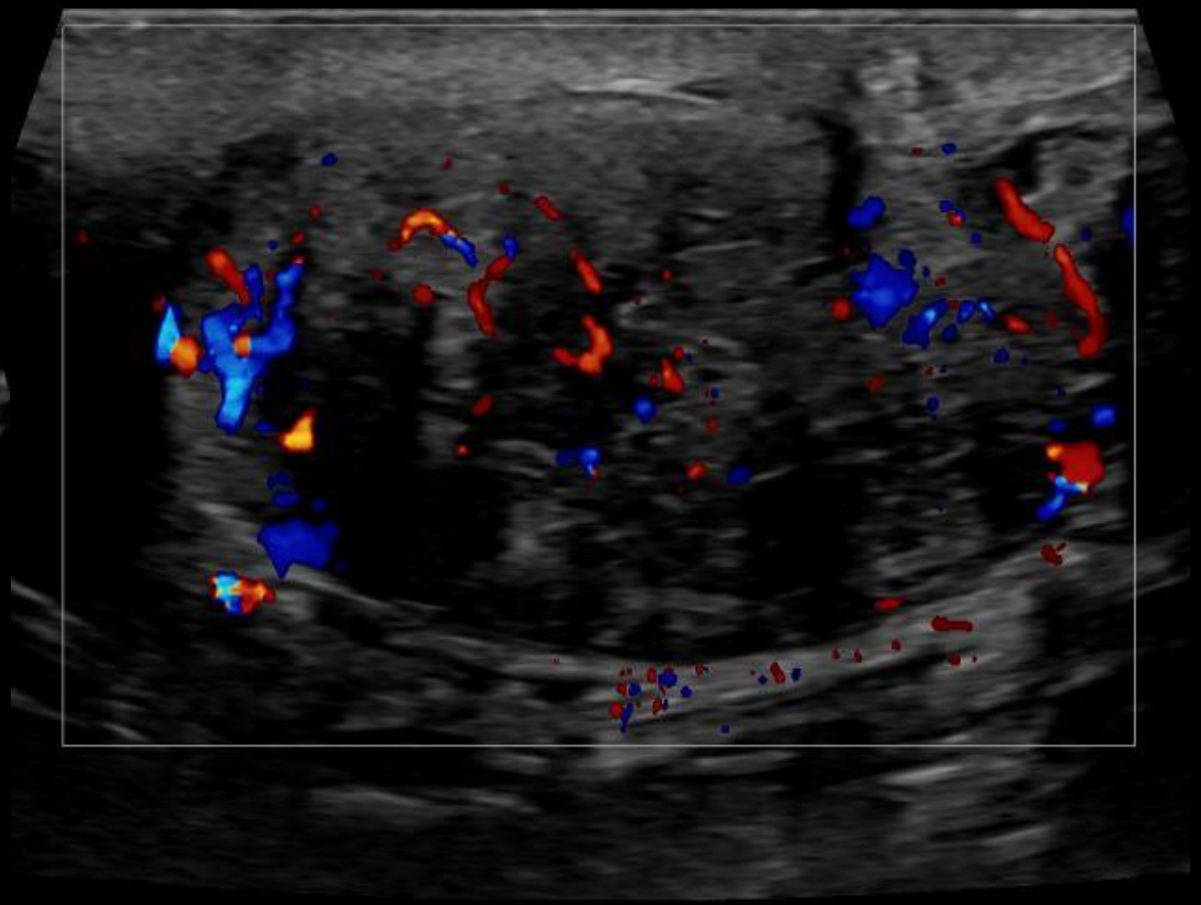
Findings (unlabeled)





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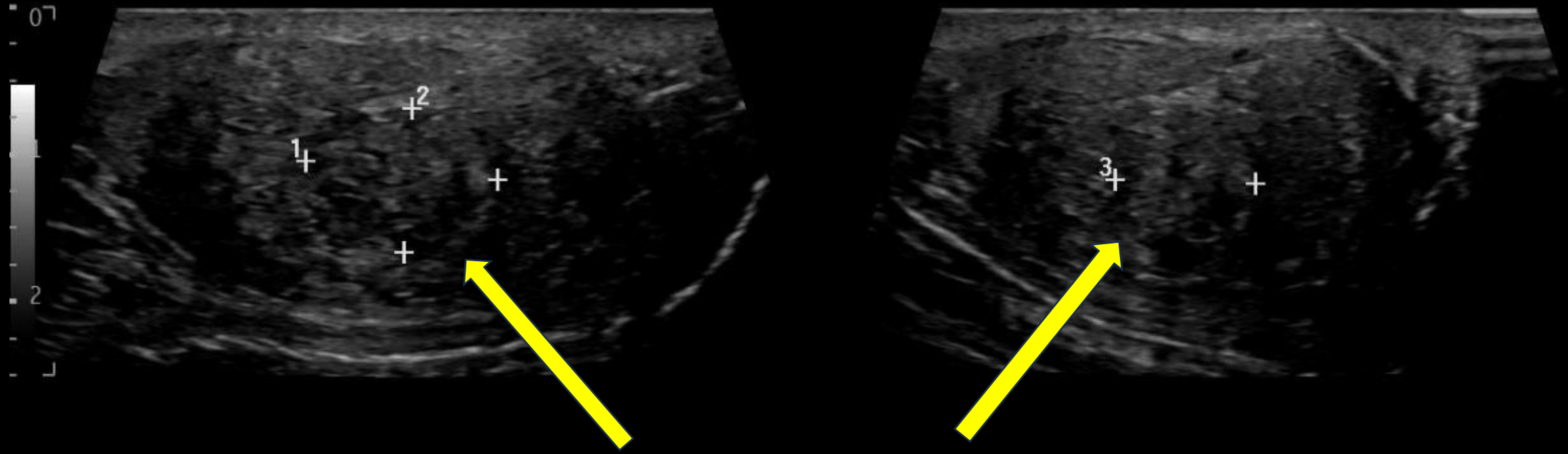
Findings: (labeled)



Nodular heterogenous echogenicity with sonographic attenuation

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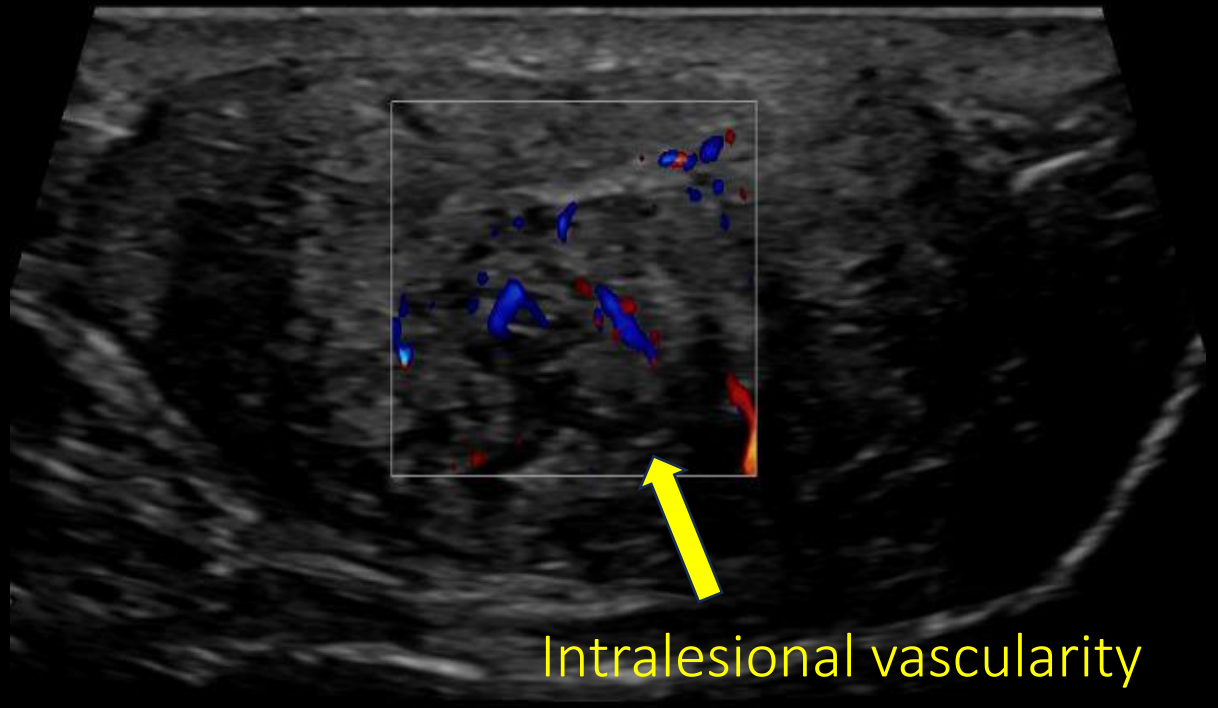
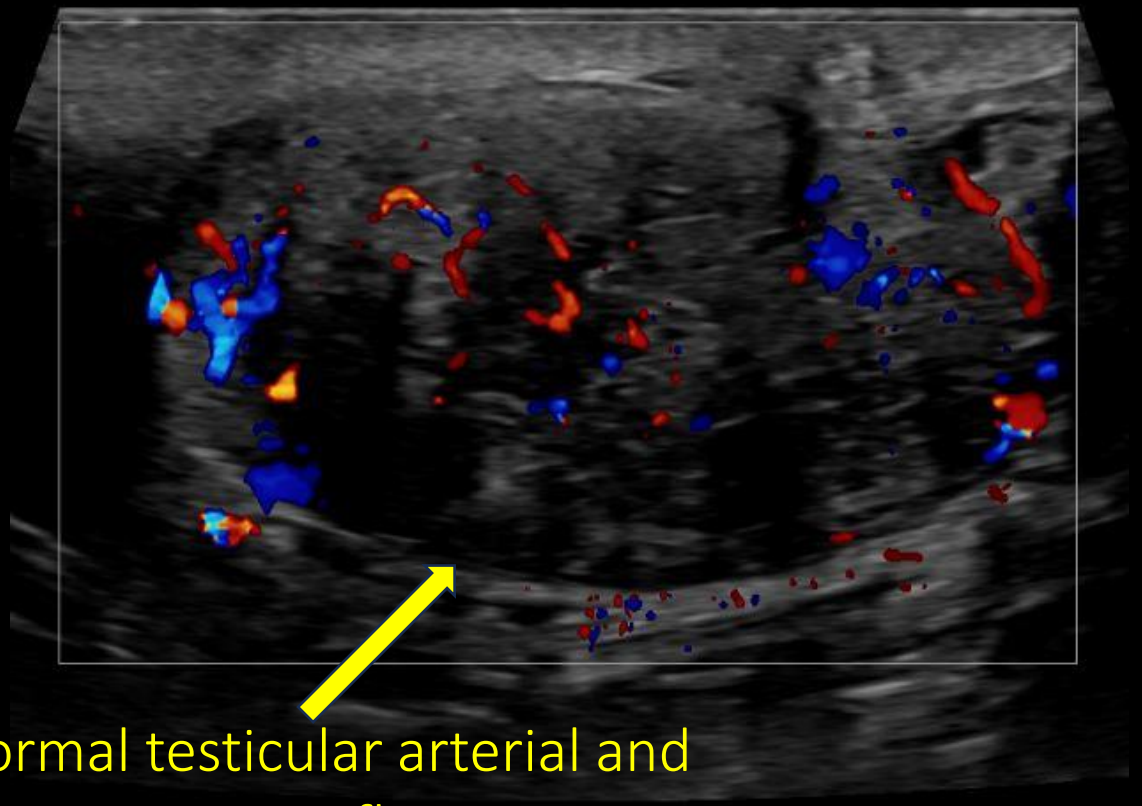
Findings: (labeled)





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Final Diagnosis:

Bilateral testicular adrenal rests

Case Discussion

- During early gestation, cells from the coelomic epithelium differentiate into steroid-producing cells of the adrenal cortex and gonads. Testicular adrenal rest tumor (TARTs) cells originate from adrenal precursor cells that migrate with gonadal cells around the eighth week of fetal development and retain ACTH responsiveness[1]
- TARTs are usually diagnosed in adulthood and are less common in children, but have been observed in boys as young as 4 years[2]

Case Discussion

- Glucocorticoid treatment aims to substitute low glucocorticoid levels and inhibit hypersecretion of ACTH and adrenal androgens[3]
- Insufficient treatment in this patient, to this point, likely explains the
  - Low levels of Cortisol, LH, and FSH
  - Elevated levels of Hydroxyprogesterone and androstenedione (precursor steroids)

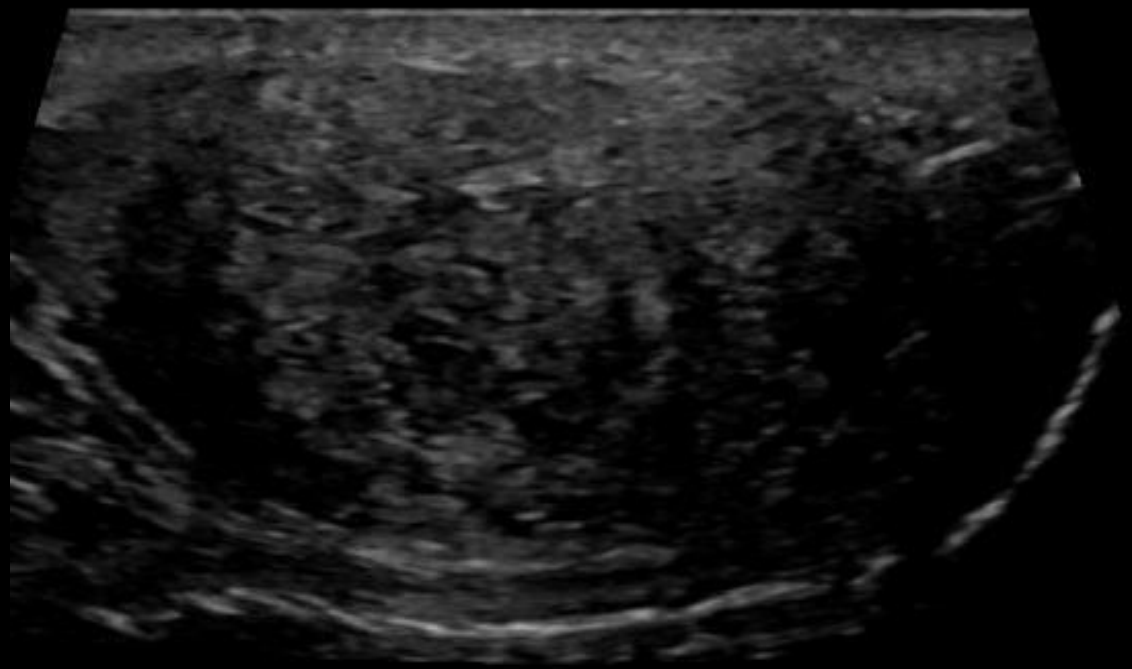
Case Discussion

- Ultrasound is the diagnostic modality of choice, and can detect small adrenal rests.[4]

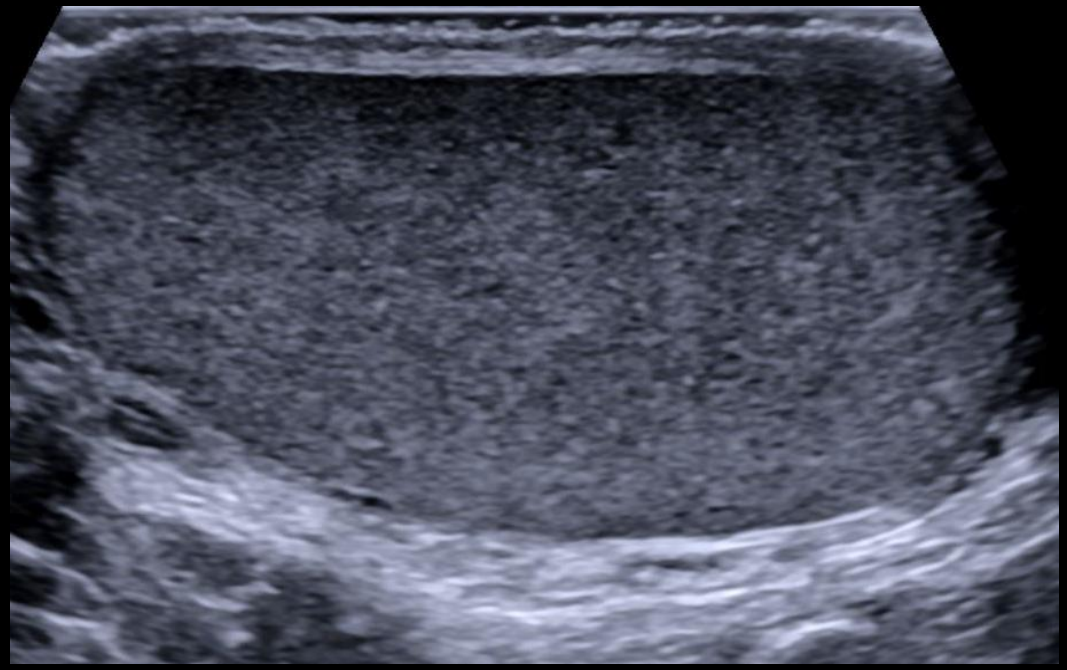
Stage	Ultrasonographic findings
Stage 1	Normal sonographic findings
Stage 2	One or more hypoechoic lesions
Stage 3	Heterogenous lesions with hyperechoic fibrous bands
Stage 4	Hyperechoic lobulated nodules with normal surrounding parenchyma
Stage 5	Hyperechoic lesions with destruction of peripheral parenchyma

Table- TART-RADS Classification[5]

Case Discussion



Heterogeneous, slightly nodular
- Stage 3/4



Normal, homogeneous

Case Discussion

- In CAH patients, TARTs are usually bilateral and reduce in size with proper medical management[6]
- Mostly benign, they can lead to obstructive azoospermia and testicular damage, causing primary gonadal failure and infertility[7]
- Annual ultrasounds are recommended to monitor growth and prevent complications like infertility[8]. If the lesion remains stable or decreases with hormonal therapy, further evaluation like biopsy or venous sampling is unnecessary[7]
- Rarely, malignant tumors can develop, such as Leydig cell tumors or seminomas[7]

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