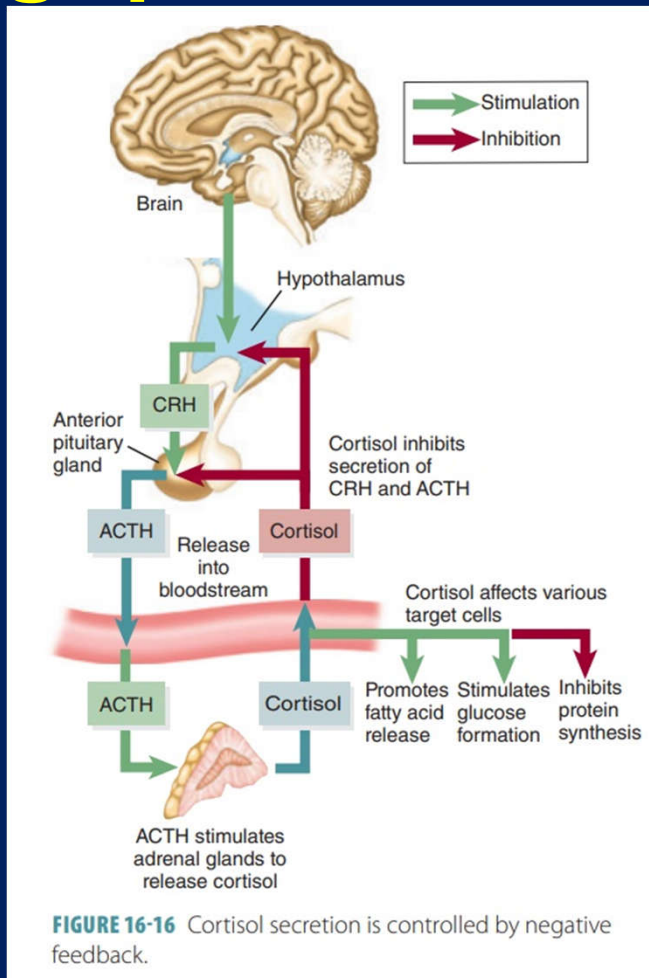




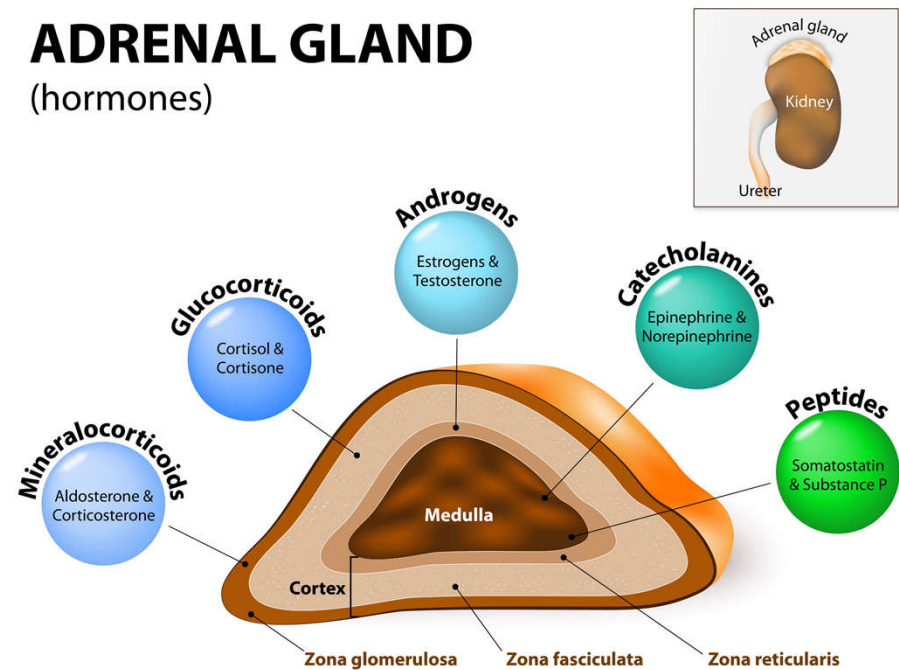
HÌNH ẢNH SIÊU ÂM U DI TÍCH THỤ ỢNG THẬN Ở TINH HOÀN

BS Lê Văn Tài
Khoa Siêu Âm MEDIC

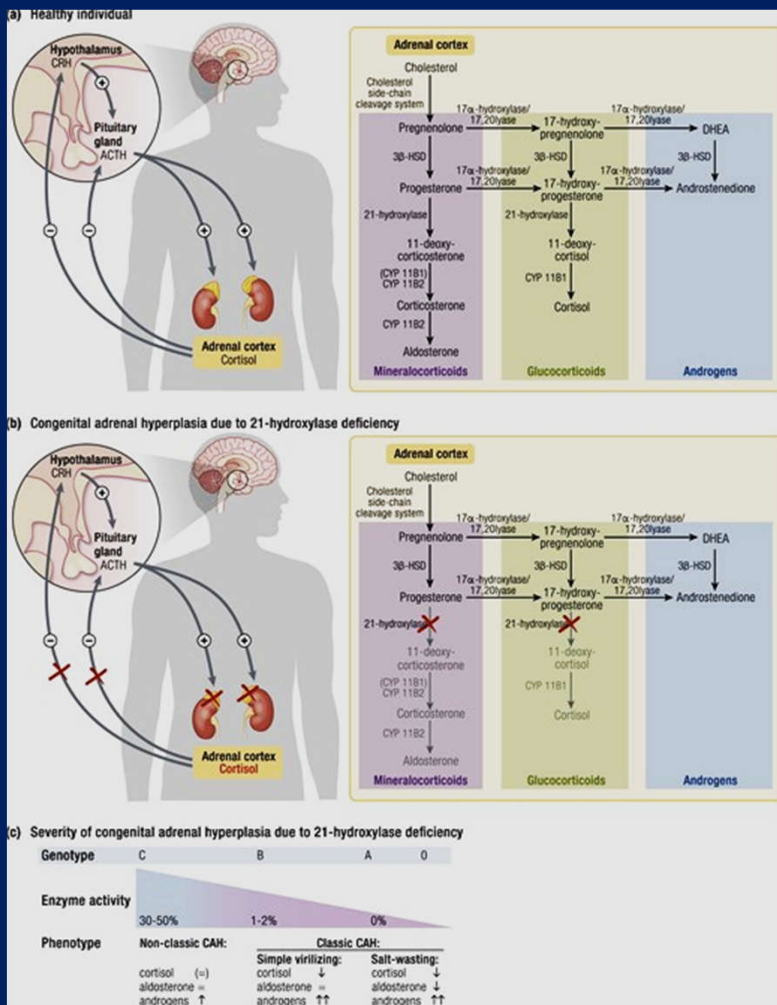
Tổng quan



ADRENAL GLAND (hormones)



Tăng sản thượng thận bẩm sinh

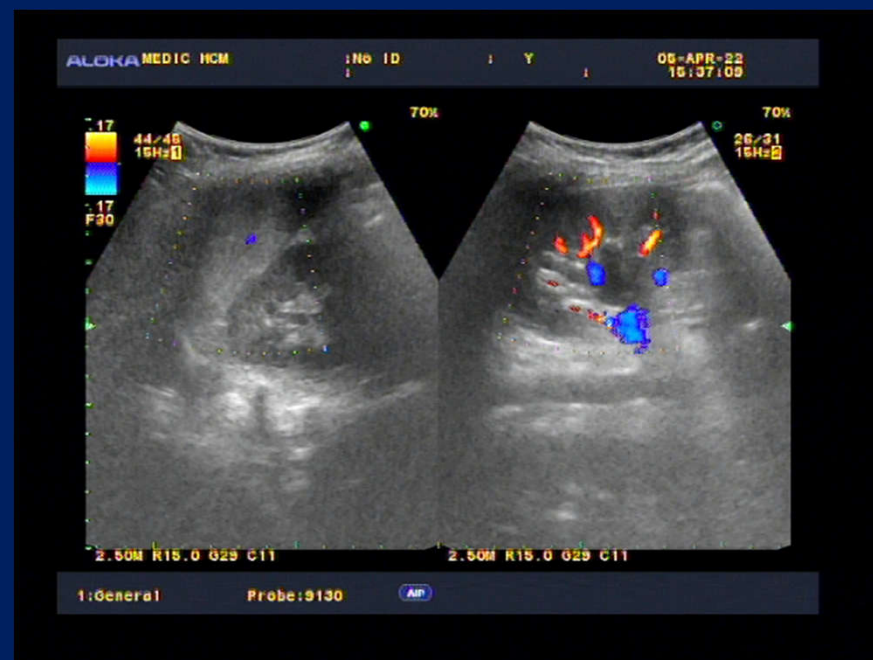
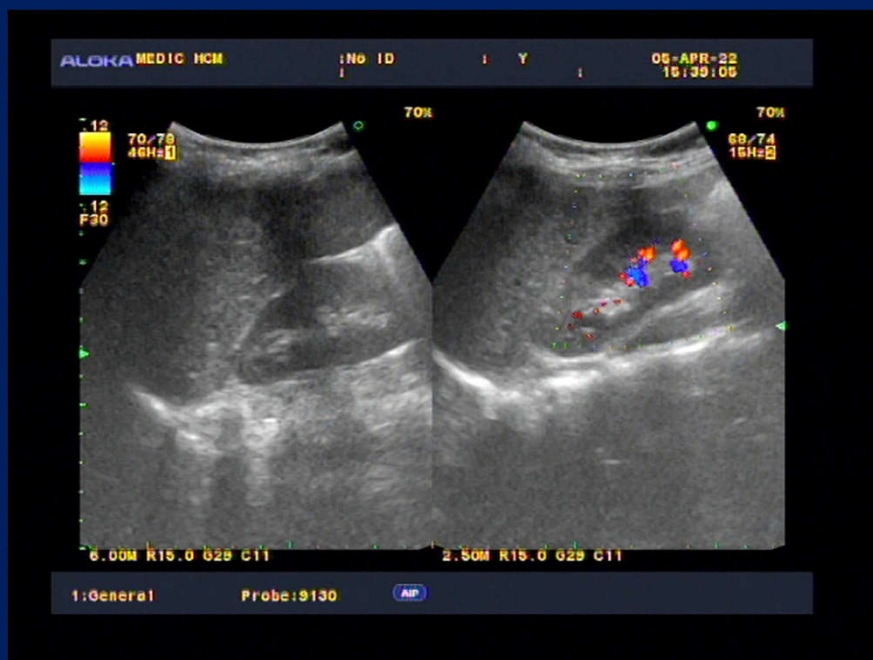


- Bệnh di truyền tính lặn trên nhiễm sắc thể thường, tỷ lệ mắc phải từ 1/10.000 đến 1/15.000. Hơn 90% do đột biến gen CYP21A2 gây thiếu enzyme 21-hydroxylase.
- Thiếu Aldosterone & cortisol, tăng androgen do ACTH kích thích liên tục tế bào thượng thận.
- Vì aldosterone tối thiểu được tiết ra, muối bị mất dẫn đến giảm natri máu, tăng kali máu & tăng hoạt tính renin huyết tương.
- Tăng androgen: bé gái buồng trứng đa nang, âm vật to, bé trai dậy thì sớm, tăng sản di tích tinh thượng thận ở tinh hoàn (tart).
- **Tăng sản thượng thận bẩm sinh cơ bản (cổ điển):** là dạng phổ biến hơn, được phân 2 nhóm nhỏ là thể mất muối và thể nam hóa.
- **Tăng sản thượng thận bẩm sinh không cơ bản (không cổ điển):** khi sinh ra trẻ vẫn khỏe mạnh bình thường, sẽ xuất hiện các triệu chứng của bệnh tăng sản thượng thận ở giai đoạn muộn.

Trường hợp thứ 1

Bé trai 7 tuổi

LS: Tăng sản thượng thận bẩm sinh đang điều trị



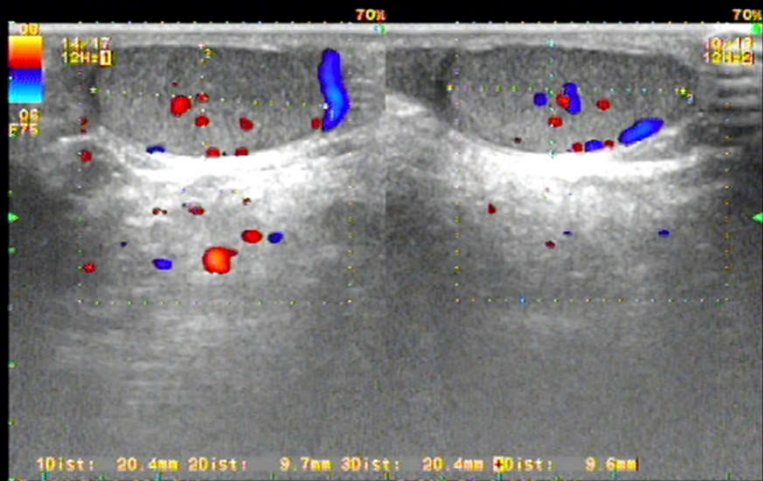
ALOKA MEDIC HCM

TRU 7M

Y

05-APR-22

10:43:26



Mark start point.

AP

SCI

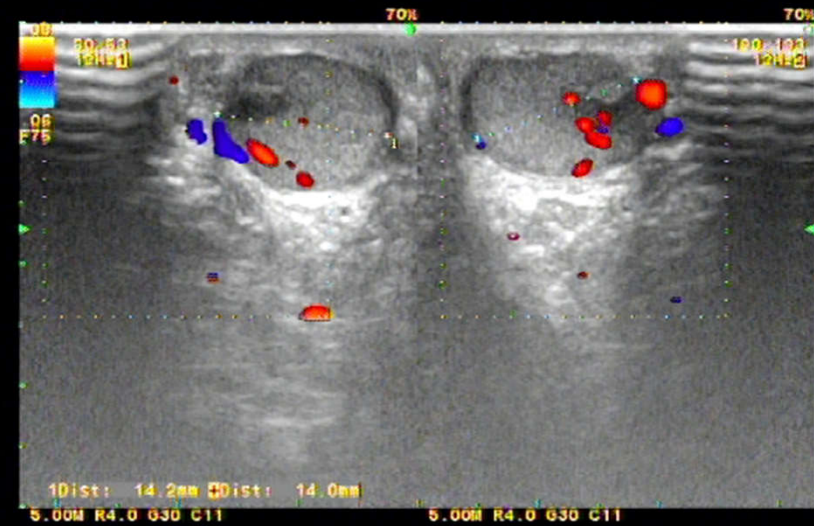
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TRU 7M

Y

05-APR-22

10:40:35



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AP

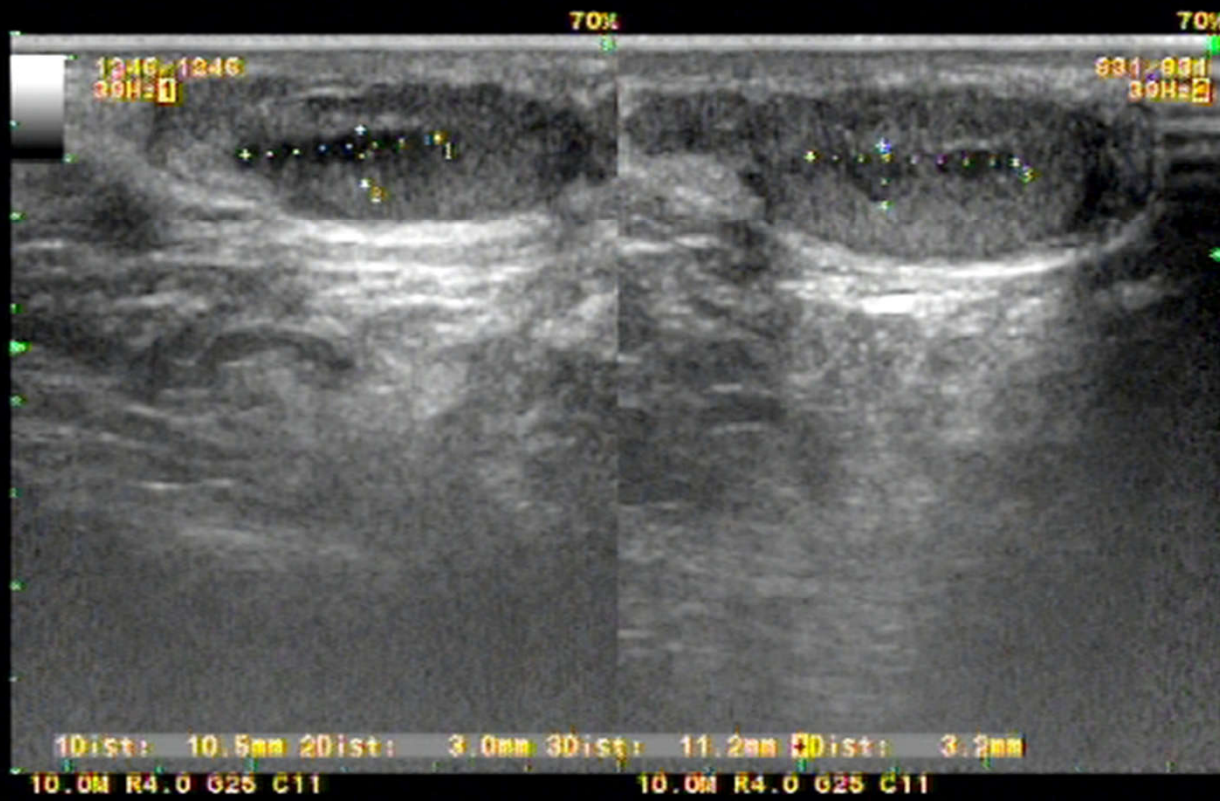
SCI

ALOKA MEDIC HCM

TRU 7M

Y

05-APR-22
15:46:38

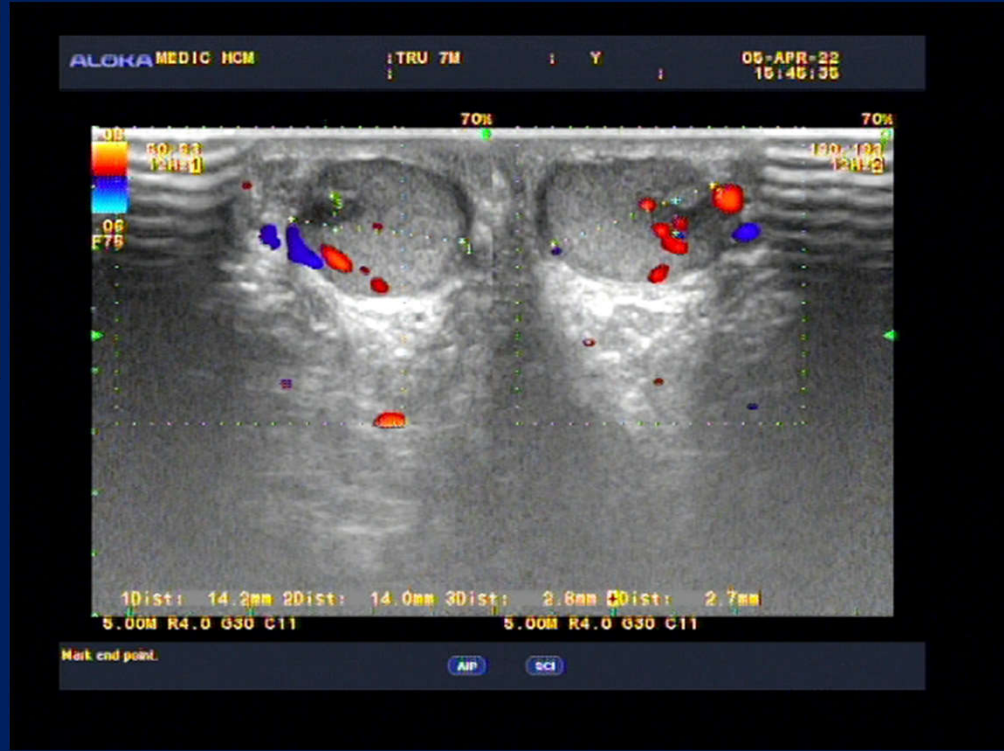
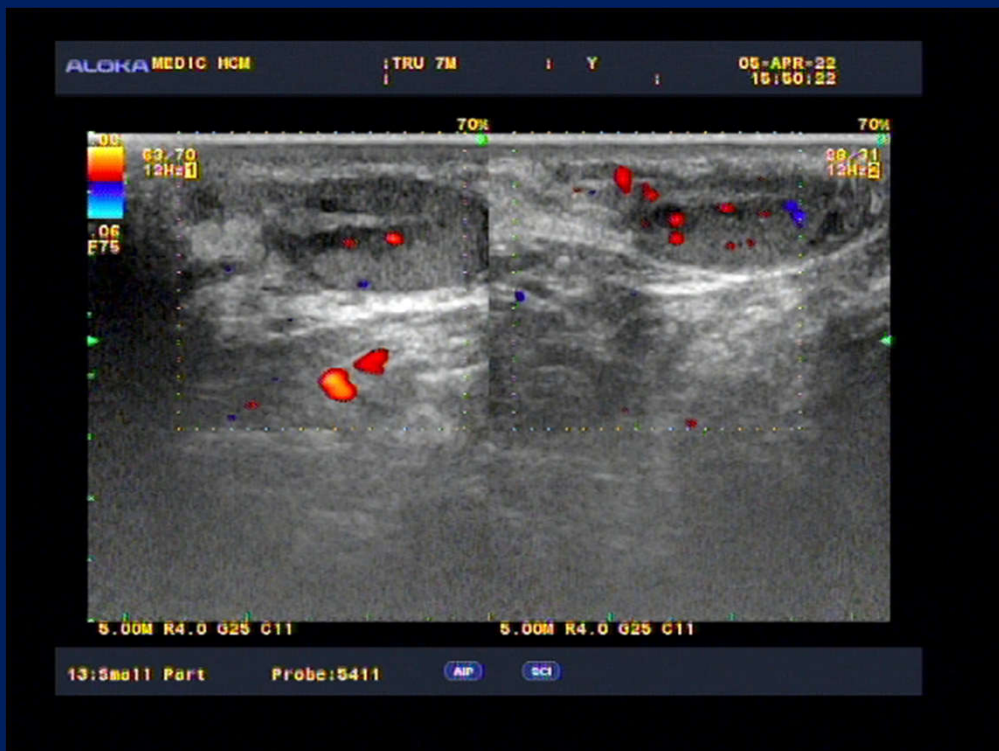


Mark end point.

AIP

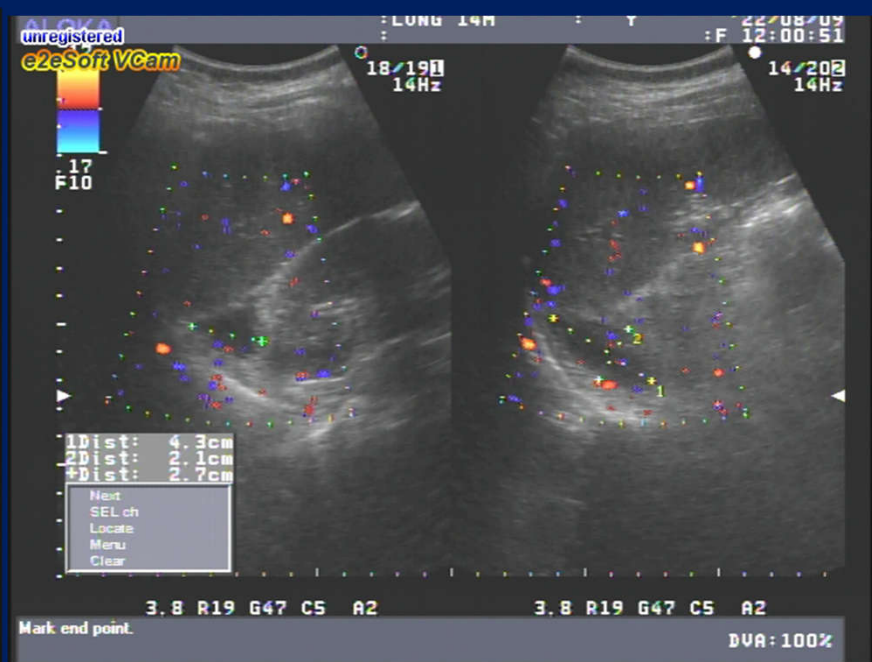
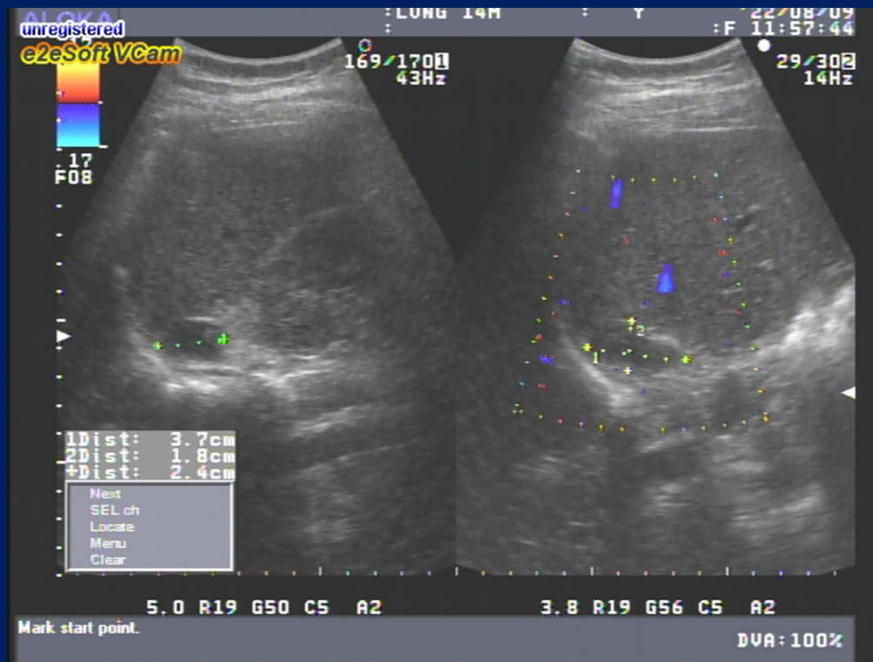
BCI

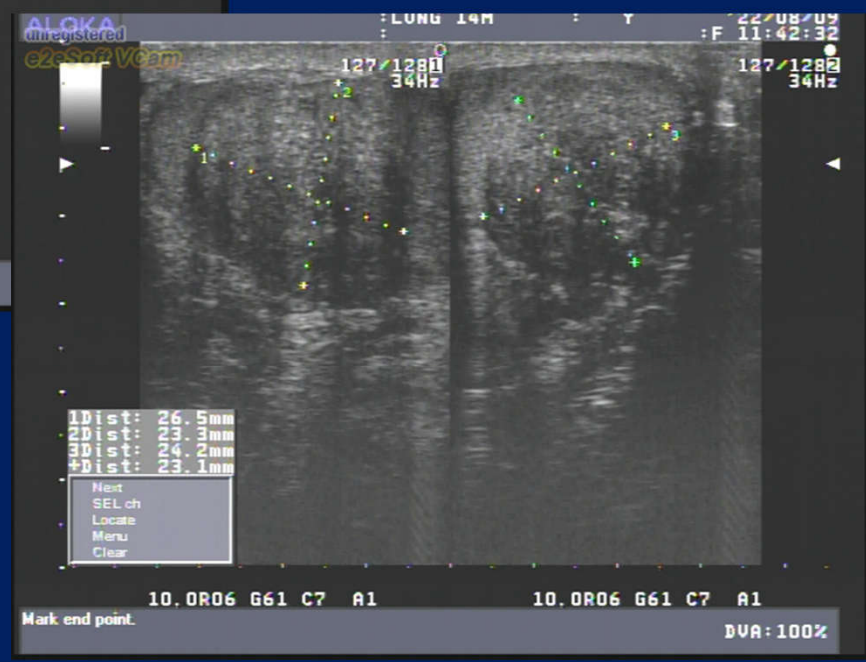
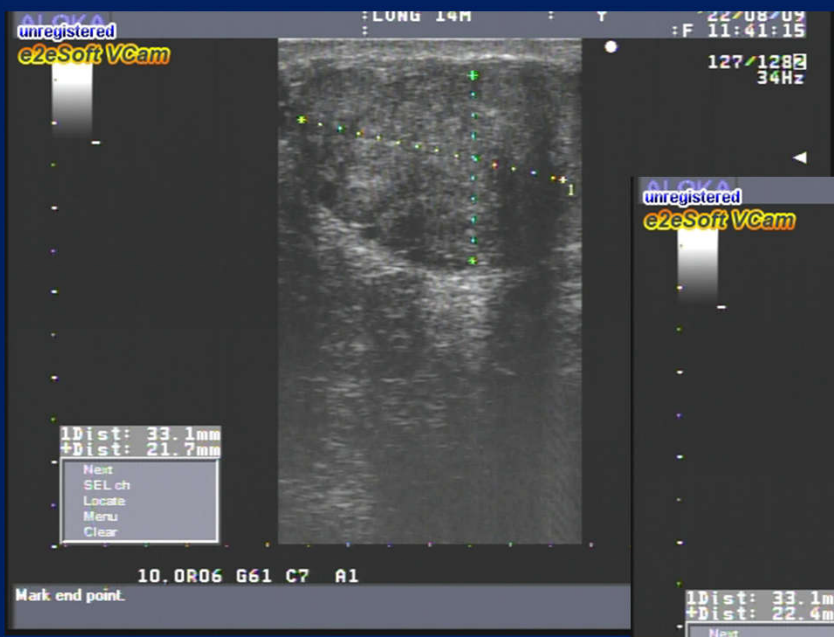


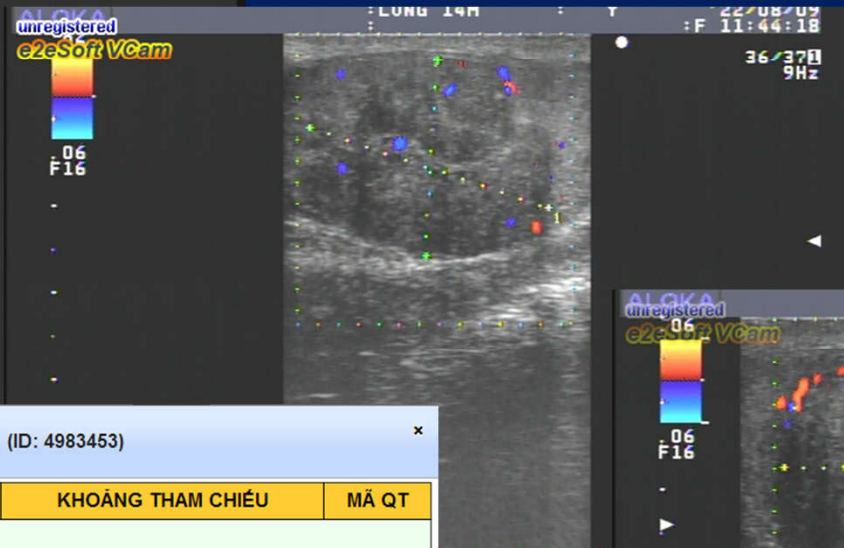
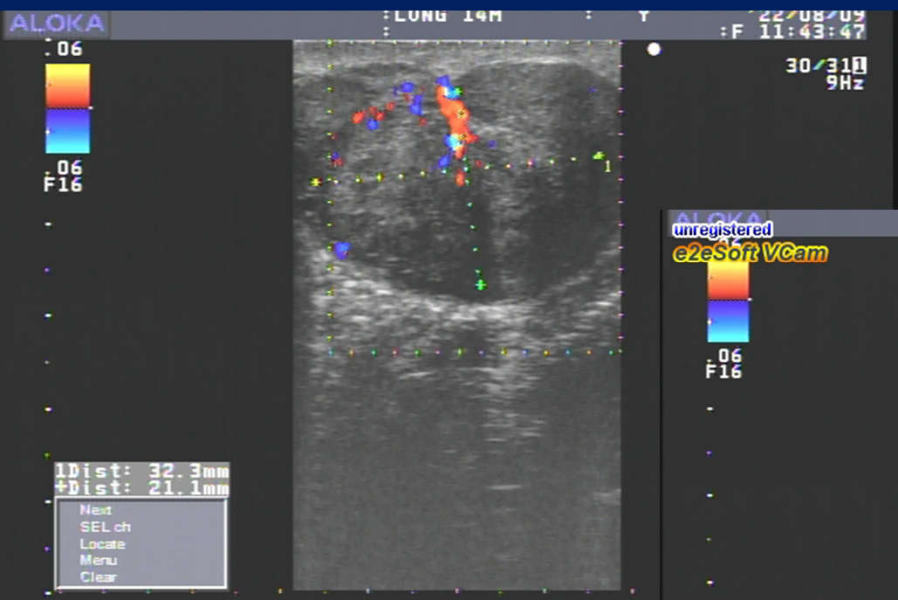


Trường hợp thứ 2

- Bệnh nhi 14 tuổi, nam
- Tăng sản thượng thận bẩm sinh điều trị từ nhỏ.





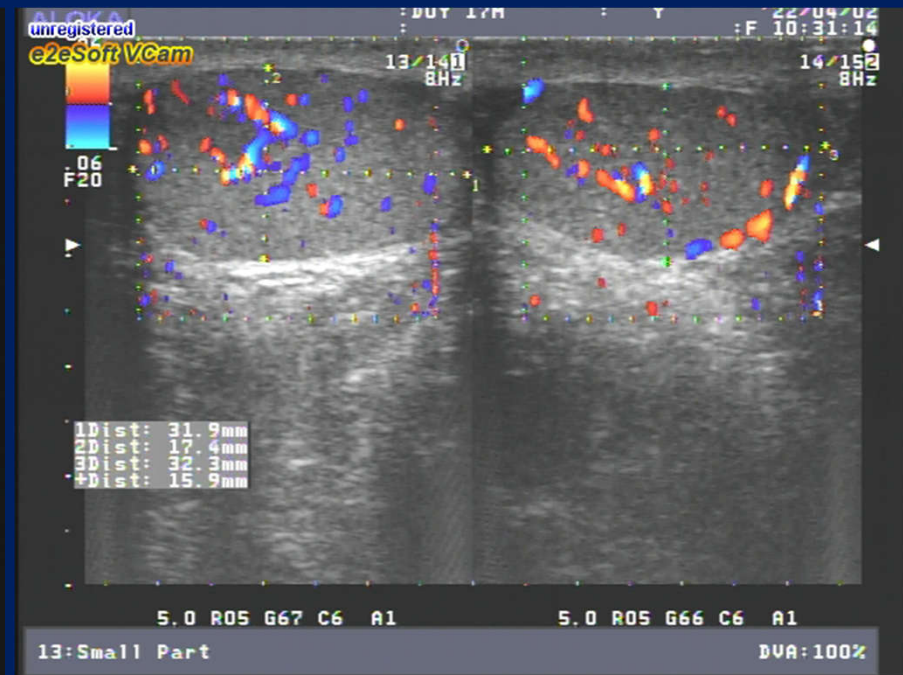


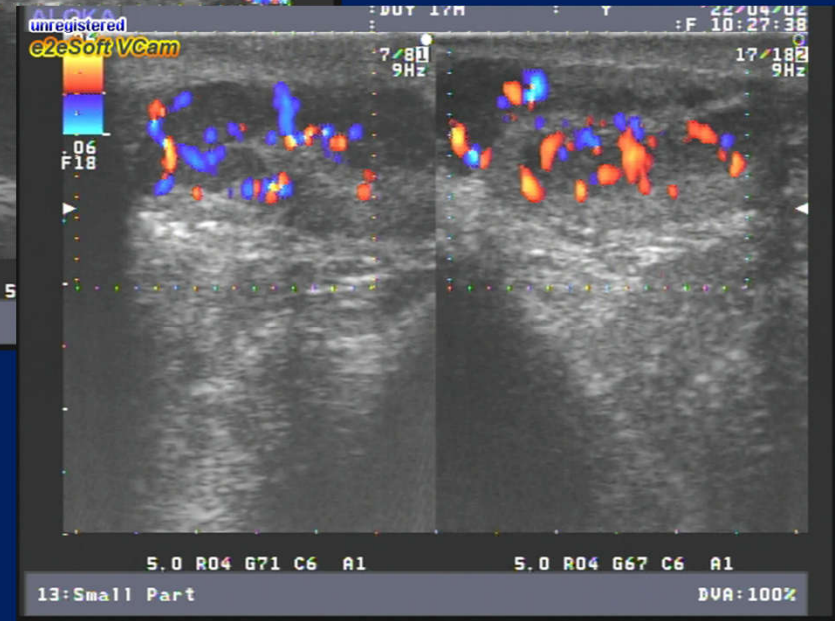
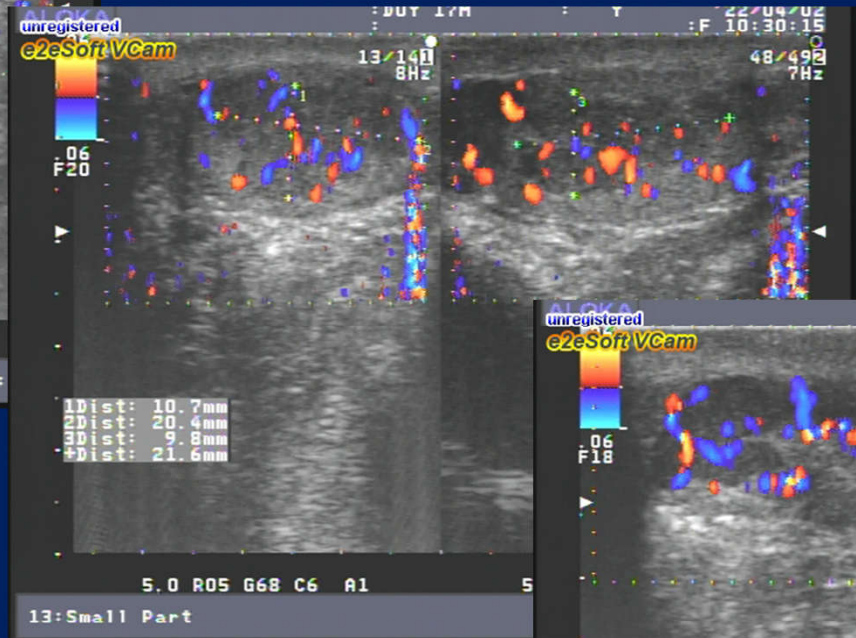
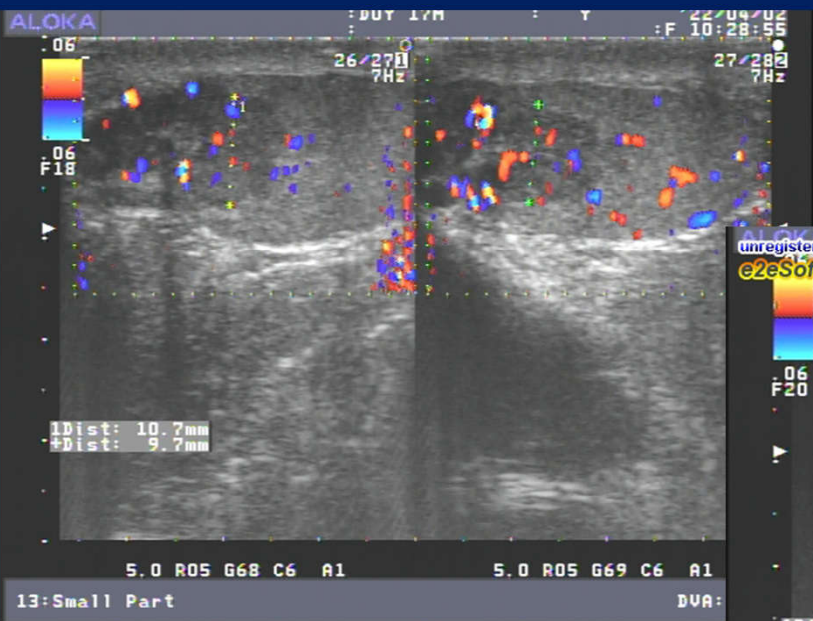
Từ ngày Đến ngày (ID: 4983453) ×

TÊN XÉT NGHIỆM	KẾT QUẢ	KHOẢNG THAM CHIẾU	MÃ QT
I. SINH HOÁ - BIOCHEMISTRY			
IONOGRAMME ² :	*		QTSH067
Na	137.3	(130 - 145 mmol/L)	
K	3.77	(3.40 - 5.1 mmol/L)	
Ca	2.35	(2.1 - 2.80 mmol/L)	
Cl	107.3	(96 - 108 mmol/L)	
II. XÉT NGHIỆM MIỄN DỊCH ELISA - ELISA TEST			
* 17-OH-Progesterone (Elisa)	31.8 ng/mL	(0.07 - 1.34 ng/mL)	
* Active Renin (Liaison):	66.15 <i>H</i>		QTMD042
	.	(Vị thể đứng: 4.4 - 46.1 µIU/mL)	
	.	(Vị thể nằm: 2.8 - 39.9 µIU/mL)	

Trường hợp thứ 3

- Bệnh nhân nam 17 tuổi
- LS: Tăng sản thượng thận bẩm sinh điều trị từ nhỏ





MEDIC - CANON 2 DR
02/04/2022

162-4786498-1W

Từ ngày 02/04/2022 Đến ngày 02/04/2022 Tìm kiếm (ID: 4786498) *

Na	138.0	(130 - 145 mmol/L)	
K	4.70	(3.40 - 5.1 mmol/L)	
Ca	2.57	(2.1 - 2.80 mmol/L)	
Cl	106.0	(96 - 108 mmol/L)	
II. XÉT NGHIỆM MIỄN DỊCH ELISA - ELISA TEST			
* 17-OH-Progesterone (Elisa)	29.28 ng/mL		
	-		
	-		
	-		
* Active Renin (Liaison):	147.2 H		QTMD042
	-	(Vị thế đứng: 4.4 - 46.1 µIU/mL)	
	-	(Vị thế nằm: 2.8 - 39.9 µIU/mL)	



Đăng ký:
02/04/2022
07:23

Xray

Bs. CKII. Hồ
Chí Trung

KTC: XQ Đo Tuổi Xương [Film]
KL: Tuổi xương > 18 tuổi

Bàn luận



- TART: là khối lành tính ở tinh hoàn, do tăng sản di tích thượng thận, xảy ra trên bệnh nhân tăng sản thượng thận bẩm sinh.
- Làm suy yếu chức năng sinh tinh & nội tiết của tinh hoàn.
- Có thể biểu hiện từ lúc nhỏ & tăng lên sau tuổi dậy thì.

- **Siêu âm:** xảy ra ở 2 tinh hoàn, khối dọc theo lưới tinh hoàn
 - Echo kém, đồng nhất hay không đồng nhất, đồng echo & không đồng nhất (heterogeneous isoechogenicity)
 - Hầu hết nhiều mạch máu, ít mạch máu (ít)
- **Chẩn đoán phân biệt:** u tinh hoàn như u tế bào Leydig (thường 1 bên).
- **Điều trị:** Liệu pháp điều trị corticosteroid thay thế kiểm hãm sự tăng sản ở bìu để giảm nguy cơ vô sinh sau này.



JOURNAL ARTICLE

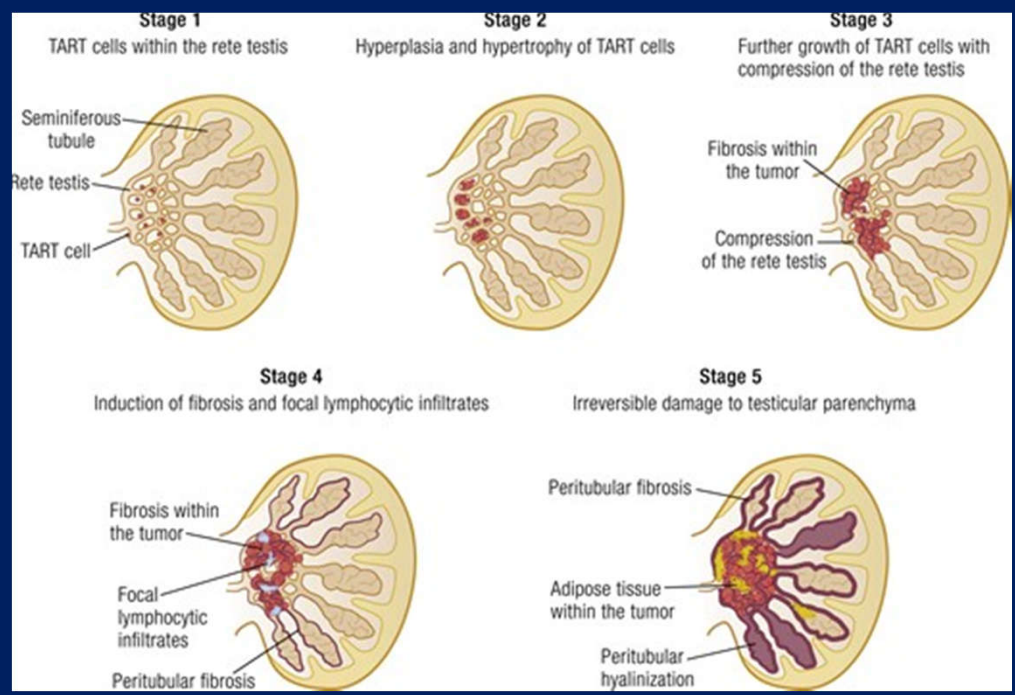
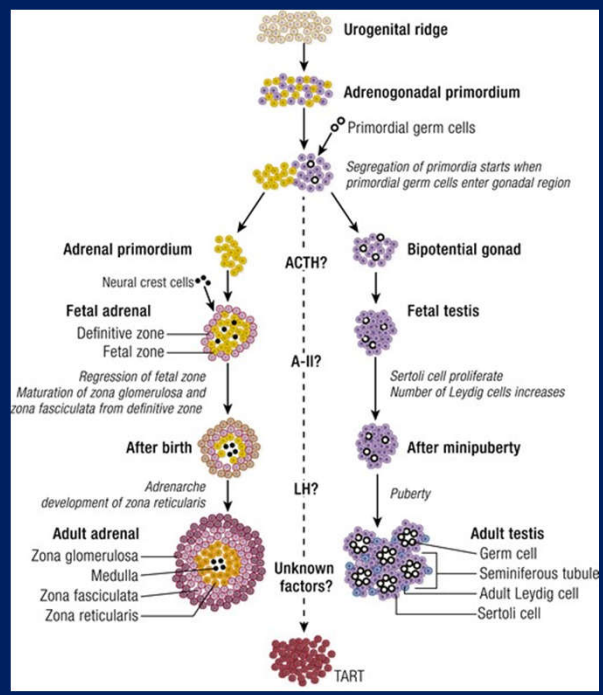
Testicular Adrenal Rest Tumors: Current Insights on Prevalence, Characteristics, Origin, and Treatment

Manon Engels, Paul N Span, Antonius E van Herwaarden, Fred C G J Sweep, Nike M M L Stikkelbroeck, Hedi L Claahsen-van der Grinten

Endocrine Reviews, Volume 40, Issue 4, August 2019, Pages 973-987, <https://doi.org/10.1210/er.2018-00258>

Published: 18 March 2019 Article history

READ A THEMATIC ISSUE ON WOMEN'S HEALTH FREE TO ACCESS ONLINE



The High Prevalence of Testicular Adrenal Rest Tumors in Adult Men With Congenital Adrenal Hyperplasia Is Correlated With ACTH Levels

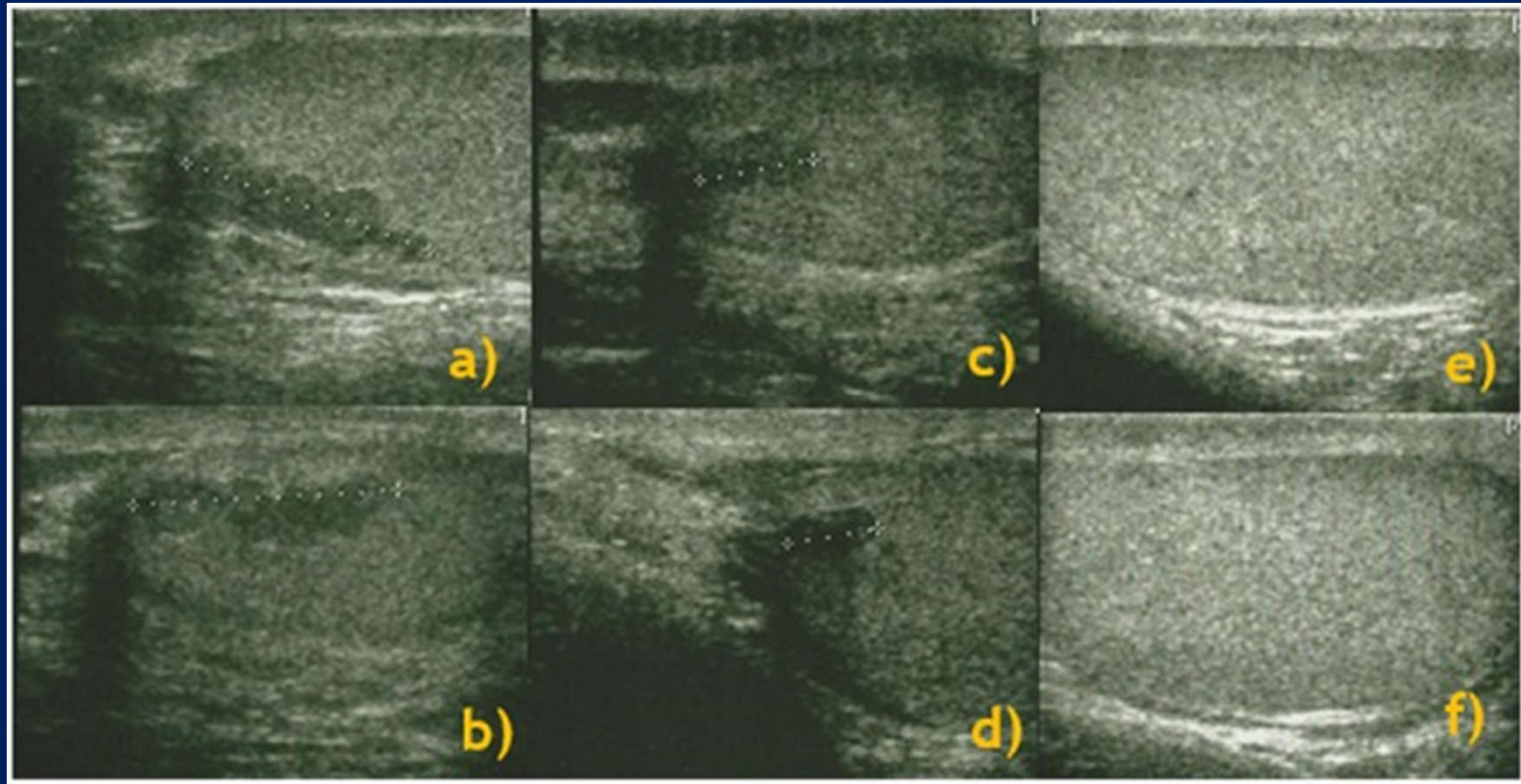
Rossella Mazzilli,^{1,2,*} Antonio Stigliano,² Michele Delfino,¹ Soraya Olana,¹ Virginia Zamponi,^{1,2} Cristina Iorio,^{1,2} Giuseppe Defeudis,³ Danilo Cimadomo,⁴ Vincenzo Toscano,² and Fernando Mazzilli^{1,2}

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Results: Of the 25 patients (mean \pm SD age, 32.2 ± 7.5 years), T-ARTs were detected by US in 14 (56.0%) patients. The mean \pm SD diameter of the lesions was 13.2 ± 6.8 mm. Among 3 (21.4%) patients, the lesions were observed in one testis, whereas both testes were affected in the remaining 11 (78.6%) patients. The lesions were hypoechoic in 12 (85.7%) patients and hyperechoic in 2 (14.3%). Plasma ACTH and 17-hydroxyprogesterone (17-OHP) levels were significantly higher in patients with T-ART than in patients without lesions (319.4 ± 307.0 pg/ml and 12.4 ± 2.7 ng/ml vs. 33.5 ± 10.7 pg/ml and 8.2 ± 1.8 ng/ml, respectively; $p < 0.01$). The mean values of sperm concentration and motility were significantly lower in patients with T-ART than in patients without lesions ($12.1 \pm 12.4 \times 10^6$ cells/ml and $18.4 \pm 11.1\%$ vs. $41.5 \pm 23.2 \times 10^6$ cells/ml and $30.8 \pm 15.4\%$, respectively; $p < 0.05$). Logistic regression analysis showed ACTH level as a significant predictor of T-ART ($p < 0.05$). In patients with T-ART, the dose of hydrocortisone was increased by ~ 25 – 30% , while the fludrocortisone treatment remained unchanged. After 6 months of steroid treatment, patients underwent US and hormonal evaluation. Disappearance and a reduction in T-ART were observed in 6 (42.9%) and 5 (35.7%) patients, respectively; a reduction in ACTH levels (from 319.4 ± 307.0 to 48.1 ± 5.1 pg/ml; $p < 0.01$) was reported. A significant correlation between ACTH level reduction and T-ART diameter reduction was observed ($p < 0.5$; $r = 0.55$).

Conclusions: T-ARTs were detected in 56% of patients with CAH and were associated with impaired semen parameters. However, these lesions are potentially reversible, as demonstrated by the disappearance/reduction after adjustment of cortisone therapy and by the reduction in plasma ACTH level. Our study supports the importance of periodic US evaluation and maintenance of plasma ACTH levels within the normal range in men with CAH.



- Longitudinal study: testis ultrasound performed at baseline [right (a) and left (b) testes], and 3 months [right (c) and left (d) testes] and 6 months [right (e) and left (f) testes] after cortisone therapy and a consequent improvement in ACTH levels.

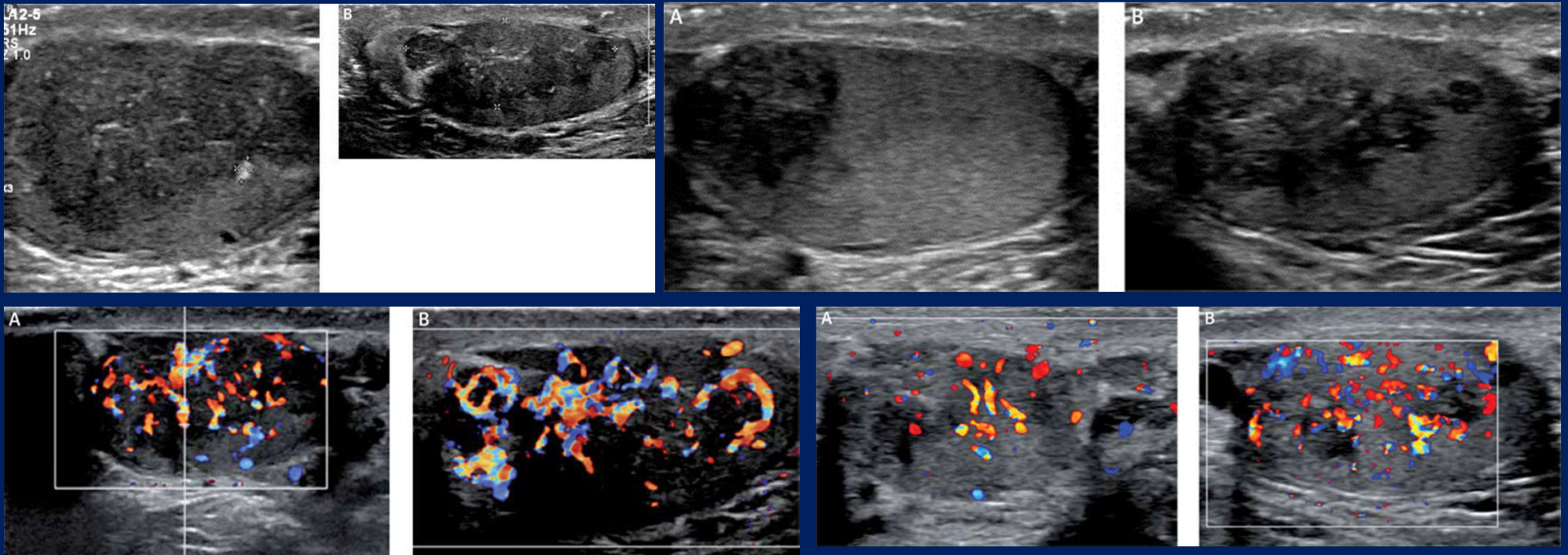
Testicular adrenal rest tumors diagnosed on ultrasound with a history of congenital adrenal hyperplasia and medication non-compliance

[Tariq Rashid](#),¹ [Andrzej Jedynak](#),¹ [Pierre-Yves Sonke](#),¹ [Irene Weiss](#),² and [Wilbert S. Aronow](#)³

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A 33-year-old man presented to the endocrinology department with a chief complaint of leg cramps. The patient was diagnosed with congenital adrenal hyperplasia due to 21-OHD at age 2 weeks. He had been receiving steroid supplementation for the majority of his life, but admitted to poor compliance with medications and medical care for about 3 years mostly due to his tenuous health insurance situation. Four days before his appointment he developed leg cramps after vigorous exercise. He was married for 2 years and recently he and his wife had been trying to conceive, without success. He had no complaints of fatigue or sexual dysfunction. On physical examination he appeared healthy, well masculinized, but had slightly small, firm testes. Laboratory evaluation returned with very high levels of 17-hydroxyprogesterone and androstenedione, low morning cortisol, and low normal testosterone. He was started on dexamethasone 1 mg at bedtime and fludrocortisone daily and a scrotal ultrasound was ordered. He returned to the endocrinology clinic after 3 months with a 15-pound weight gain. He had not yet undergone the scrotal ultrasound. Because of the weight gain his glucocorticoid was changed to hydrocortisone three times daily with doses of 10 mg, 5 mg, and 5 mg. Six months later, on his next follow-up visit, a scrotal ultrasound was reordered.

- Initial sonographic evaluation (November 2017) showed bilateral hypoechoic heterogeneous vascular intratesticular masses (A – right testicle, B – left testicle) with irregular borders extending to the epididymal heads estimated to occupy approximately 50% of the testicular parenchyma Doppler interrogation showed extensive vascularity within both masses (A – right testicle, B – left testicle).



Six-month follow-up sonographic evaluation of both masses (April 2018) showed that glucocorticoid up-titration after initial ultrasound revealed that the testicular masses had minimally decreased in size (A – right testicle, B – left testicle). Internal vascularity of 6-month sonographic evaluation shows decreased vascularity (A – right testicle, B – left testicle)

Sonographic features of the testicular adrenal rests tumors in patients with congenital adrenal hyperplasia: a single-center experience and literature review

Li Ma, Yu Xia, Linlin Wang, Ruifeng Liu, Xuepei Huang, Tiantian Ye, Li Zhang, Qingli Zhu, Jianchu Li & Yuxin Jiang 

Orphanet Journal of Rare Diseases 14, Article number: 242 (2019) | [Cite this article](#)



Results

A total of 8 patients aged between 4 to 27 years old were enrolled. 7 of 8 (87.5%) patients exhibited bilateral testicular lesions. The sizes of the testicular lesions were between 0.18 ml to 5.68 ml, and all showed a clear boundary. 10/15 (66.7%) lesions were homogeneously hypoechoic, 4/15 (26.7%) were heterogeneously iso-hypoechoic, and 1/15 (6.7%) were homogeneously isoechoic. 10/15 (66.7%) lesions were hyper-vascular. The longitudinal follow-up of 5 patients showed testicular lesions changed in terms of size, echogenicity, and vascularity after steroid treatment. A potential correlation may exist between ACTH levels and tumor size ($p = 0.066$). From the literature review, 100/123 (81%) patients got bilateral lesions, and 95% of them were located near the mediastinum. 80/103 (78%) lesions exhibited a clear boundary, and predominant lesions (74%) were hypoechoic. Vascularity was with great diversity. Seventy-nine lesions of 44 patients were followed-up by scrotal ultrasound, among which 29 (37%) remained unchanged, 29(37%) shrank, and 21(27%) disappeared.

Conclusions

Key sonographic characteristics of TART are: resembled lesions on both testes, located near the mediastinum, clear boundary, and changed in size or echogenicity after steroid treatment. These features can help radiologists to make an accurate diagnosis of TART.



- Ultrasound features of TART. This is a 13-year-old CAH patient. Palpable testicular nodules were detected through physical examination and a scrotal ultrasound was therefore performed. The scrotal gray-scale ultrasound (a) and color Doppler ultrasound (b) showed homogeneous hypoechoic lesions with clear boundary on both testes with marked vascularity. Follow-up gray-scale (c) and color Doppler (d) ultrasound examination was performed after a 6-month steroid treatment, which showed a remarkable decrease in lesion size and vascularity

Kết luận



- Nên siêu âm sớm cho trẻ em cường thượng thận bẩm sinh để tìm TART là cấu trúc echo kém nhiều mạch máu ở 2 tinh hoàn, cạnh trung thất tinh hoàn.
- Siêu âm có vai trò phát hiện & theo dõi điều trị giúp ngăn chặn sự phát triển lớn ra của TART để bảo tồn mô tinh hoàn.

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1. Manon Engels, Paul N. Span, Antonius E. van Herwaarden, Fred C. G. J. Sweep, Nike M. M. L. Stikkelbroeck, and Hedi L. Claahsen-van der Grinten. Testicular Adrenal Rest Tumors: Current Insights on Prevalence, Characteristics, Origin, and Treatment. *Endocrine Reviews*, Volume 40, Issue 4, August 2019, Pages 973–987, <https://doi.org/10.1210/er.2018-00258>
2. Li Ma, Yu Xia, Linlin Wang, Ruifeng Liu, Xuepei Huang, Tiantian Ye, Li Zhang, Qingli Zhu, Jianchu Li & Yuxin Jiang. Sonographic features of the testicular adrenal rests tumors in patients with congenital adrenal hyperplasia: a single-center experience and literature review. *Orphanet Journal of Rare Diseases* volume 14, Article number: 242 (2019)
3. Rossella Mazzilli, Antonio Stigliano, Michele Delfino, Soraya Olana, Virginia Zamponi, Cristina Iorio, Giuseppe Defeudis, Danilo Cimadomo, Vincenzo Toscano, and Fernando Mazzilli. The High Prevalence of Testicular Adrenal Rest Tumors in Adult Men With Congenital Adrenal Hyperplasia Is Correlated With ACTH Levels. *Front Endocrinol (Lausanne)* 2019; 10: 335. Published online 2019 Jun 4. doi: 10.3389/fendo.2019.00335
4. Tariq Rashid, Andrzej Jedynak, Pierre-Yves Sonke, Irene Weiss, and Wilbert S. Aronowcorresponding author. Testicular adrenal rest tumors diagnosed on ultrasound with a history of congenital adrenal hyperplasia and medication non-compliance. *Arch Med Sci.* 2020; 16(6): 1501–1504. Published online 2020 Nov 2. doi: 10.5114/aoms.2020.100310

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