

TUMORI DELL'INFANZIA E ADOLESCENZA



Le dimensioni del problema

Tumore in bambini e adolescenti

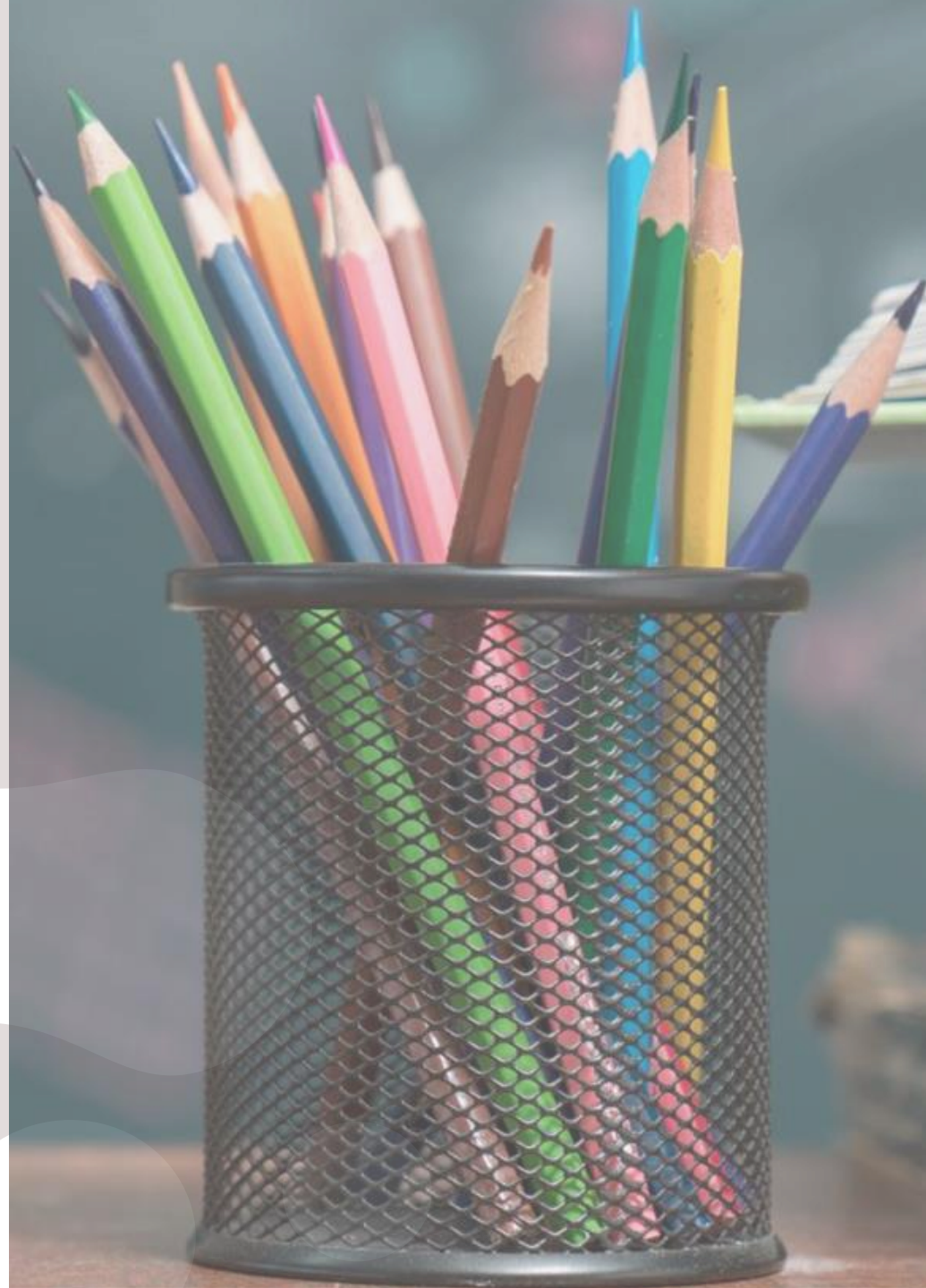


EVENTO RARO

Incidenza stabile negli ultimi anni in Italia

164 casi / milione di bambini (0-14 anni)

269 casi / milione di adolescenti (15-19 anni)



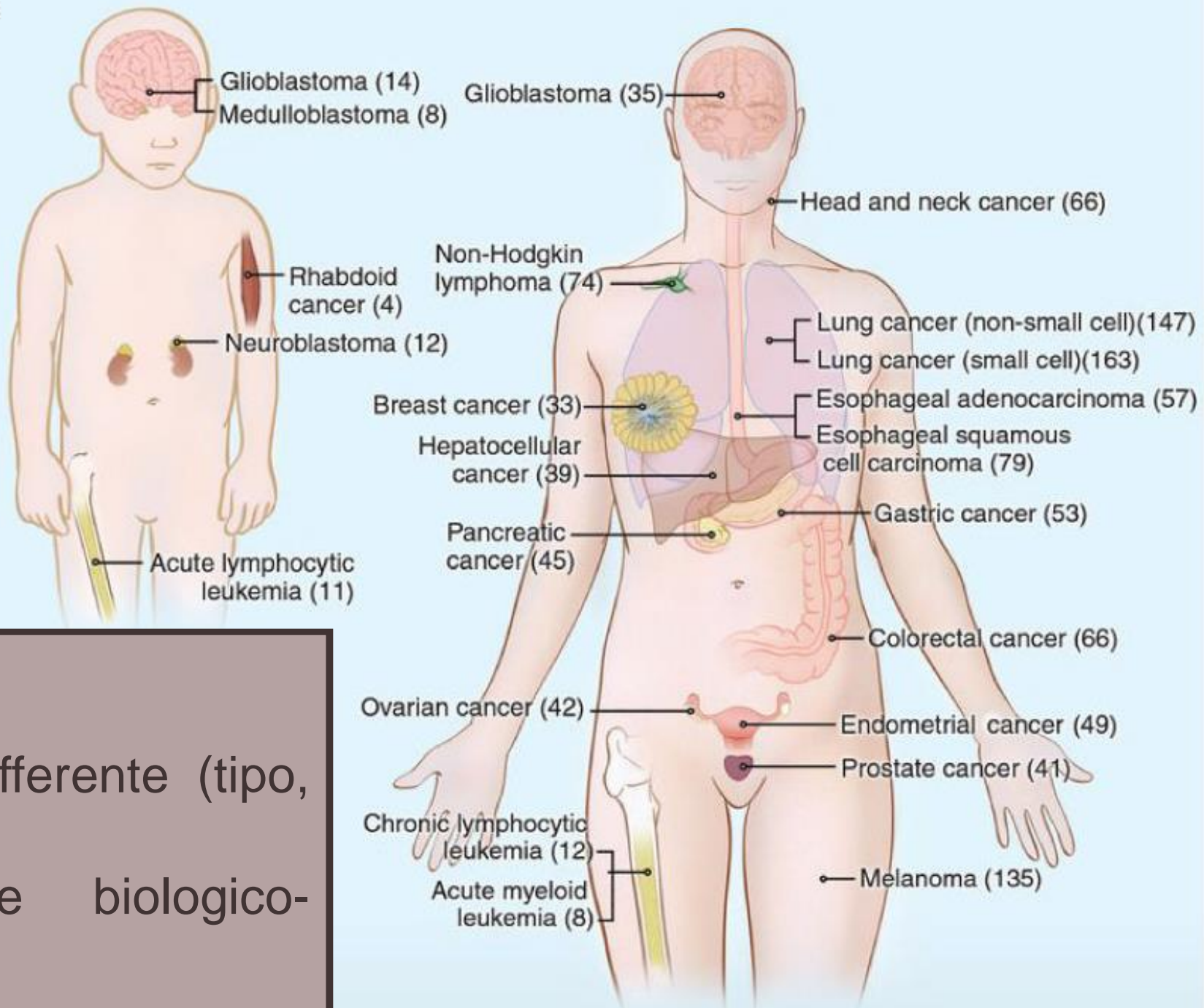
Tumore in bambini e adolescenti

EVENTO RARO

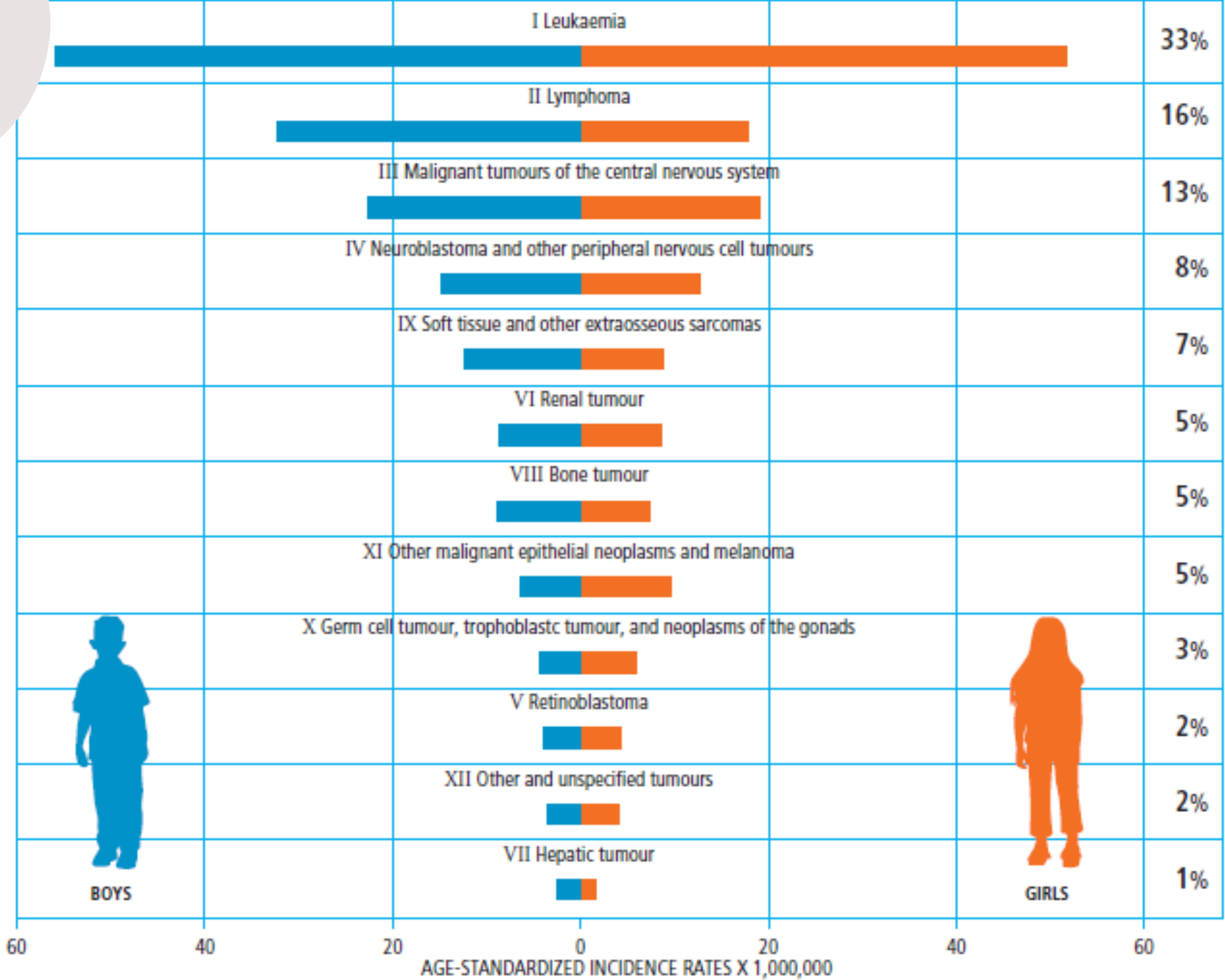
Pochi dati

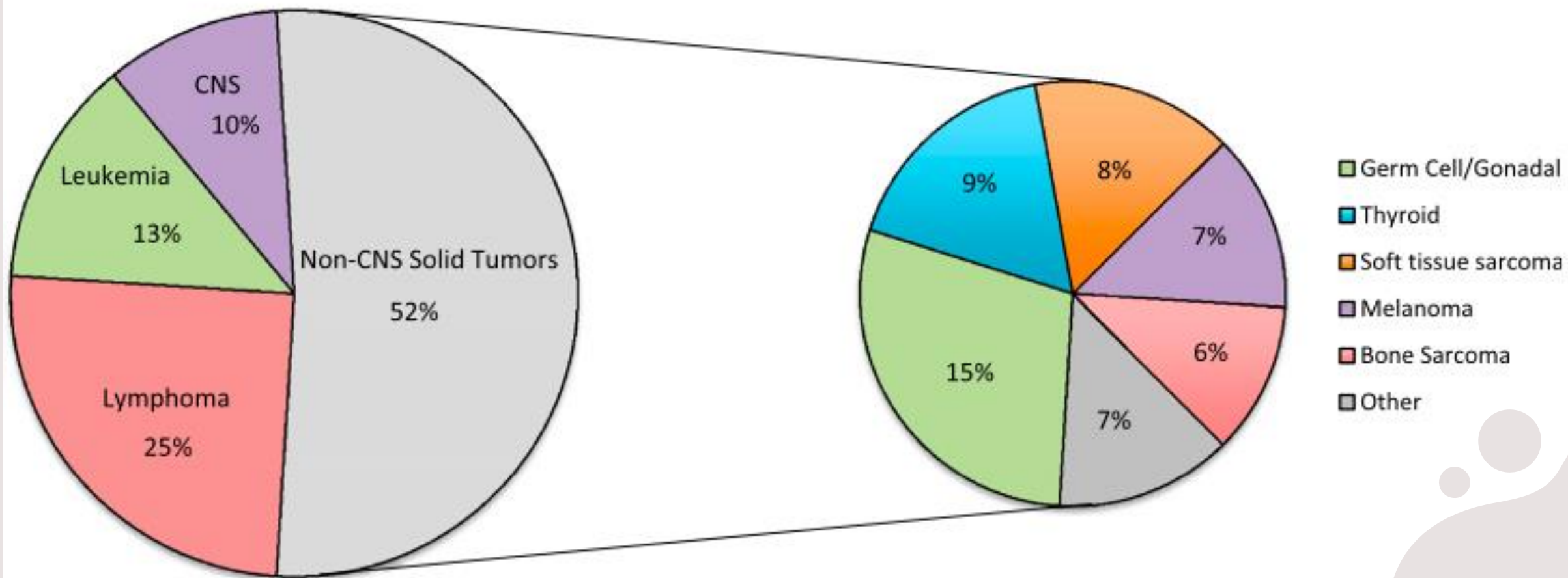
- Numero inferiore di casi
- Spettro di patologie differente (tipo, età)
- Diverse caratteristiche biologico-molecolari

A

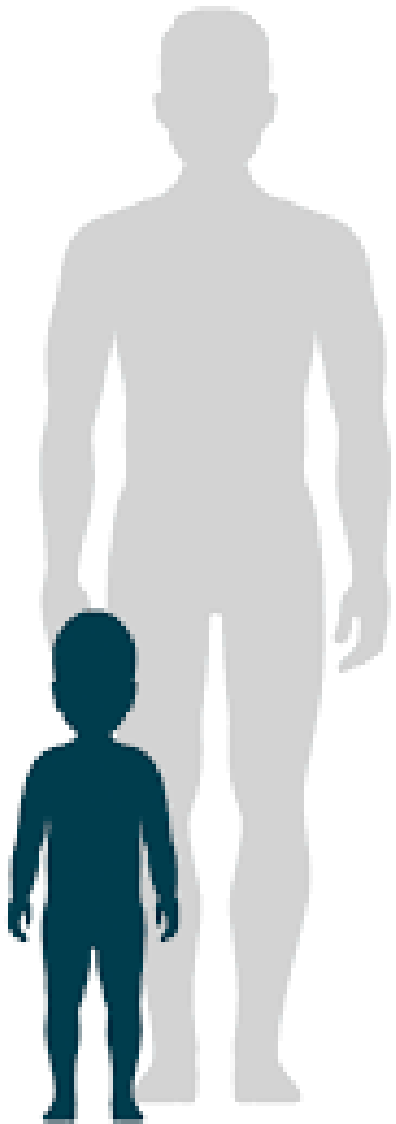


I S T O T I P I





Distribuzione istotipi tumorali in ADOLESCENTI



PROGNOSI

Possibilità di guarigione superiore rispetto ai tumori dell'adulto

Leucemie pediatriche >85%

Tumori solidi pediatrici 60-70%

Prognosi variabile a seconda di:

- Tipo di neoplasia*
- Estensione di malattia (stadio)*

DIAGNOSI

- Non facile
- Considerare il tumore come possibile diagnosi differenziale



dolore



tumefazione / gonfiore



febbre



stanchezza



dimagrimento



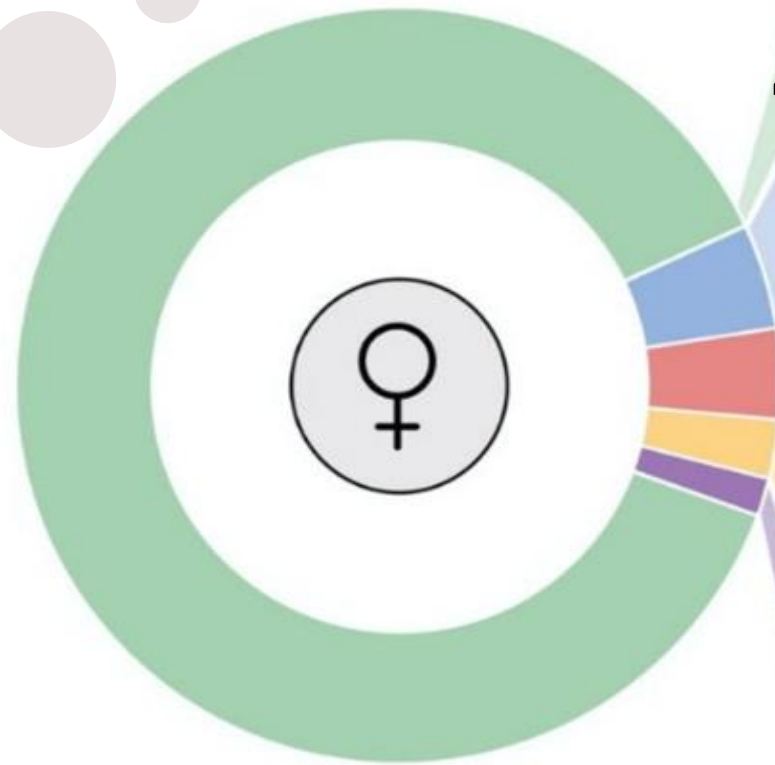
sintomi persistenti



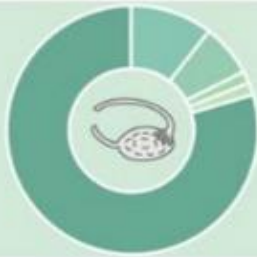
TUMORI GINECOLOGICI

4.2 % di tutti i tumori
nelle femmine ≤ 18 anni





87.5%



- Germ-cell tumor (79.6%)
- Epithelial cancer (10.6%)
- Sex-cord-stromal tumors (6.9%)
- Sarcoma (1.4%)
- Other /n.s. (1.5%)

OVARIAN T.

4.5%



- Sarcoma (69.6%)
- Germ-cell tumors (26.8%)
- Clear cell carcinoma (3.6%)

VAGINAL T.

3.9%



- Sarcoma (42.9%)
- Adenocarcinoma (34.7%)
- Squamous cell carcinoma (10.2%)
- Other / n.s. (12.2%)

CERVICAL T.

2.5%



- Sarcoma (34.4%)
- Adenocarcinoma (34.4%)
- Choriocarcinoma (21.9%)
- Germ-cell tumor (6.3%)
- Other / n.s. (3.1%)

UTERINE T.

1.6%

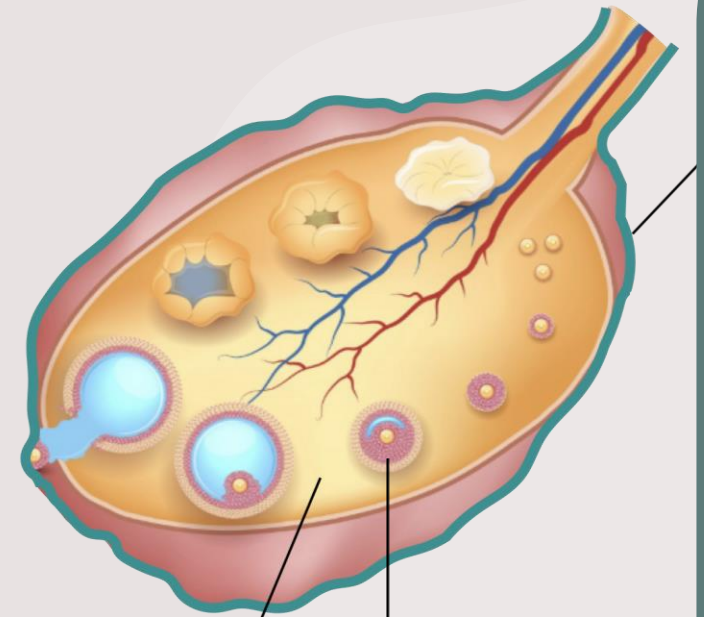


- Sarcoma (65.0%)
- Melanoma (15.0%)
- Squamous cell carcinoma (10.0%)
- Germ-cell tumor (5.0%)
- Other / n.s. (5.0%)

VULVAR T.

OVARIAN T.

80%



Germ cell tumors

Stromal tumors

**5 year survival
97.2%**

OGCT

Germ cells

Neoplastic transformation

MATURE

EMBRYONAL

BENIGN
✓ Mature cystic teratoma
✓ Struma ovarii

MALIGNANT
✓ Squamous carcinoma
✓ Carcinoid
✓ Thyroid cancer

UNDIFFERENTIATED
Disgerminoma

SOMATIC
Immature teratoma

PRIMITIVE
Embryonal carcinoma

TROPHOBLASTIC
Choriocarcinoma

EXTRAEMBRYONAL
Yolk sac tumor



TERATOMA MATURO CISTICO

- *80% Monolaterale*
- *Spesso peduncolato*
- *Dimensioni variabili (da 1 a 30-40 cm)*
- *Growth rate 1.8 mm/anno*

**50% delle
formazioni
annessiali
<20 aa**



*Easy
descriptors*

BD2



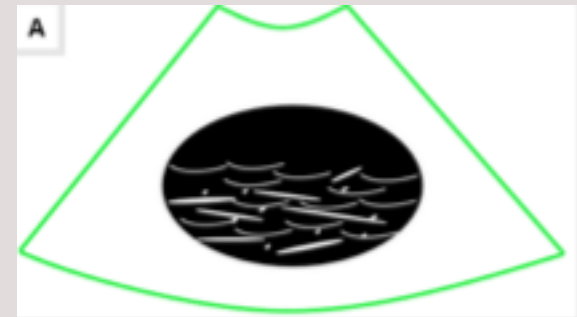
- *Formazione uniloculare*
 - *Ecogenicità mista*
 - *Shadows*
 - *Premenopausa*



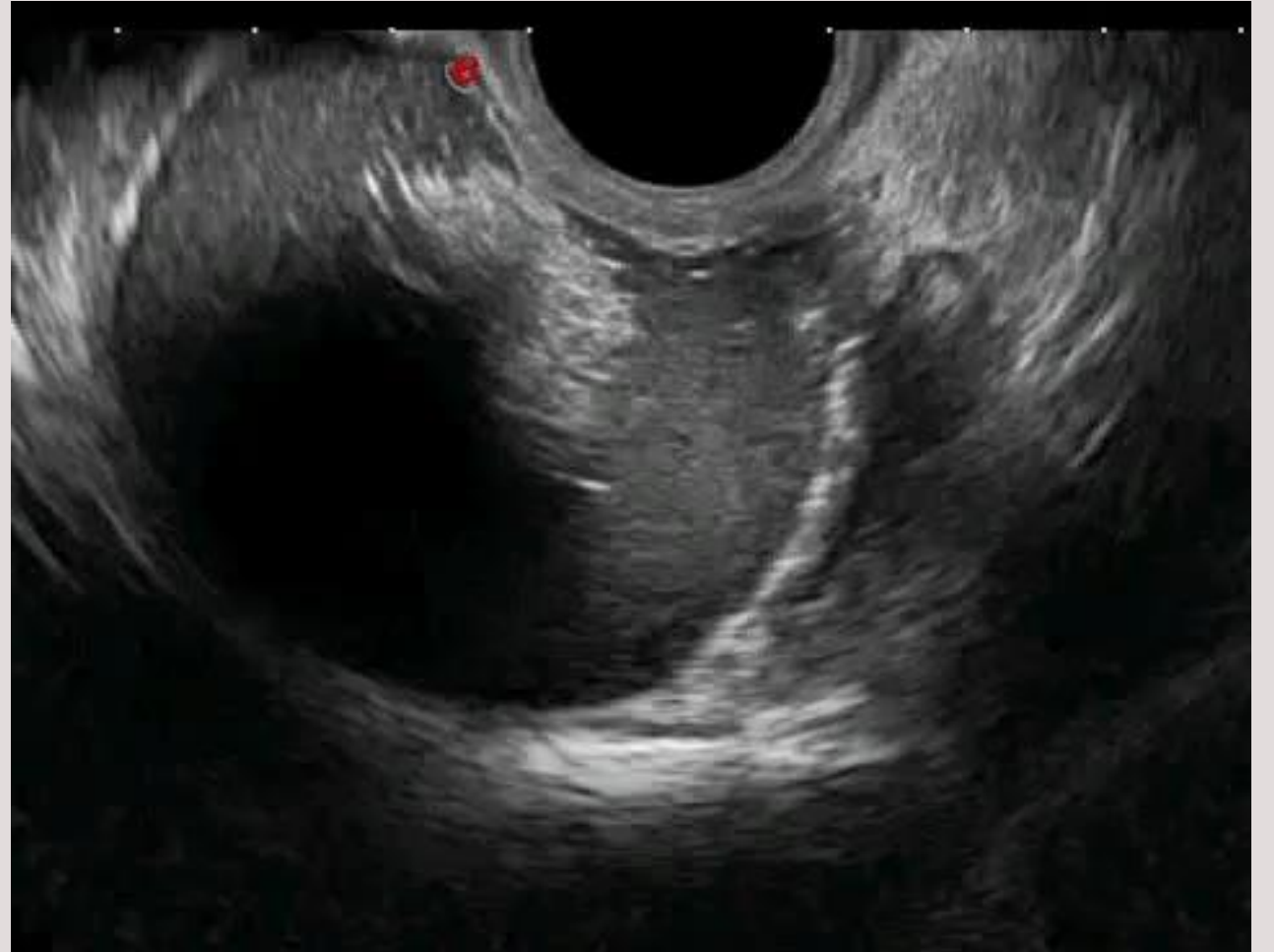
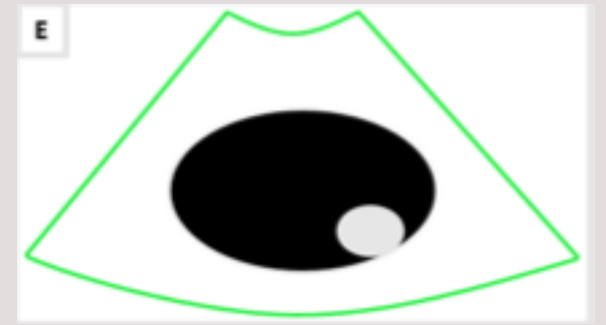
Imaging in gynecological disease: clinical and ultrasound characteristics of ovarian mature cystic teratomas

R. Heremans^{1,2}, L. Valentin^{3,4}, P. Sladkevicius³, S. Timmerman^{1,2}, F. Moro⁵, C. Van Holsbeke^{1,6}, E. Epstein⁷, A. C. Testa⁵, D. Timmerman^{1,2} and W. Froyman^{1,2*}

1. DERMOID MESH (dots & lines)



2. 'WHITE BALL'



HERAW10

0
7
14



3. FAT FLUID SIGN



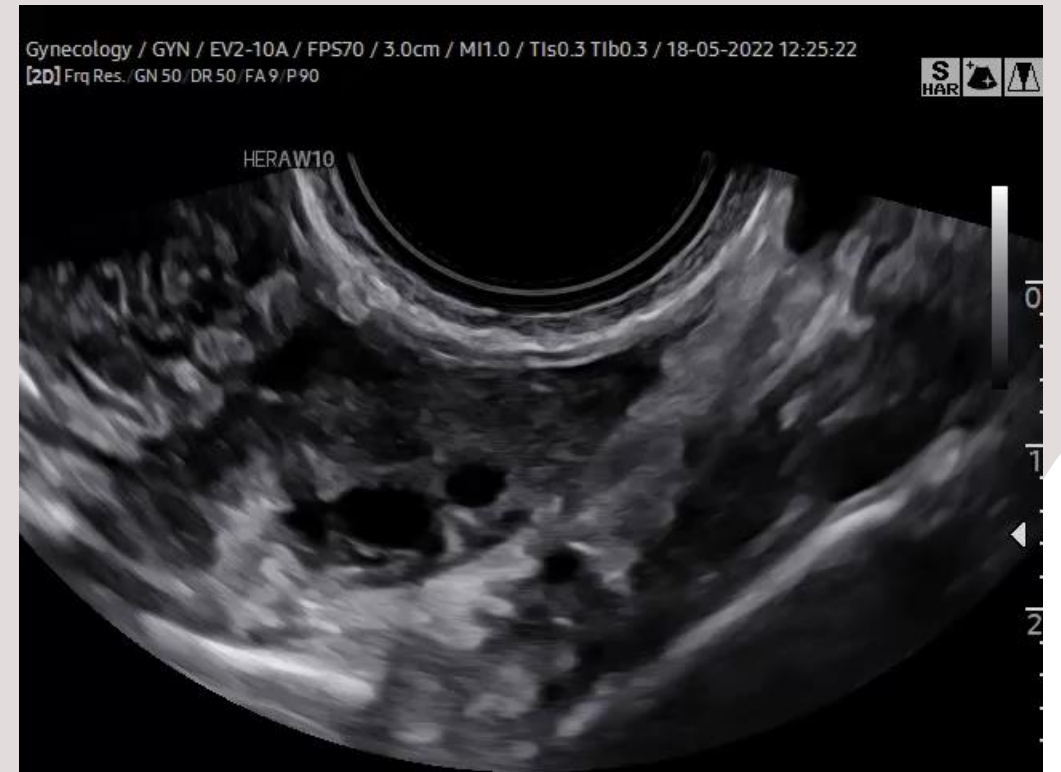
4. TIP OF THE ICEBERG SIGN



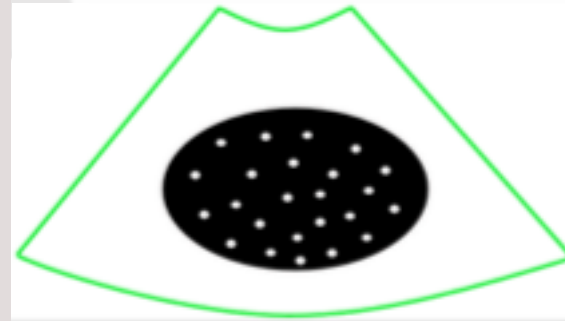
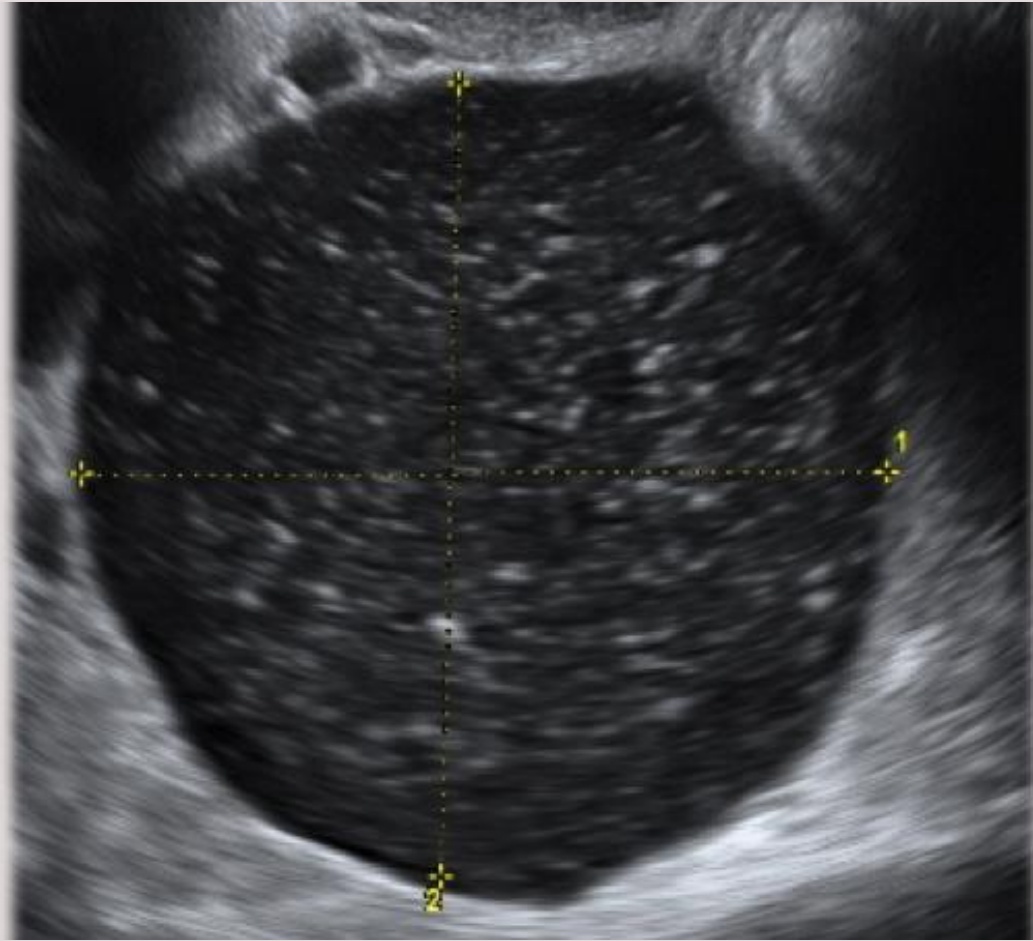


5. COTTON WOOL TUFTS

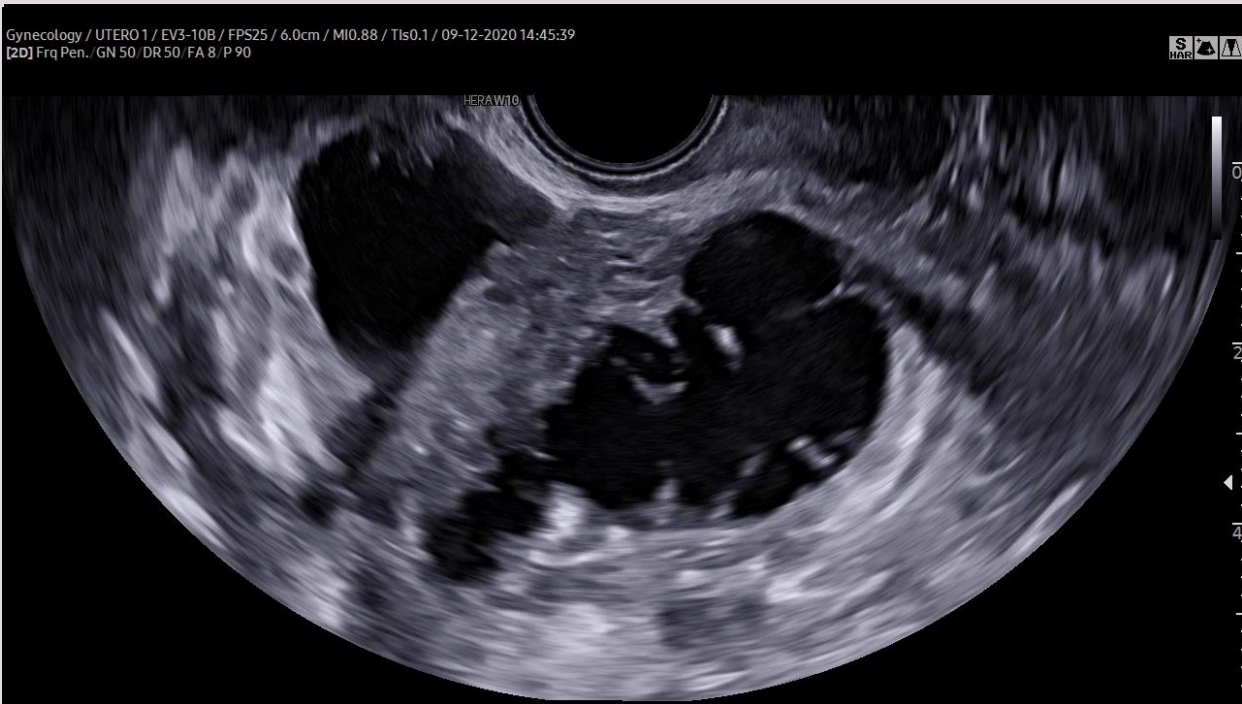
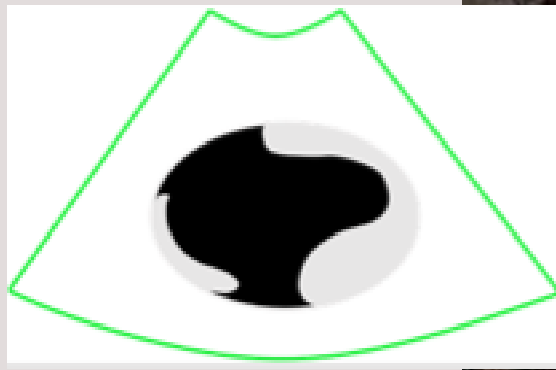
6. COMPLETELY ECHOGENIC LESION



7. STARRY NIGHT SIGN



8. MUSHROOM CUP SIGN



Gynecology / UTERO 1 / EV3-10B / FPS30 / 6.0cm / MI1.0 / TIs0.2 / 23-11-2020 11:18:04
[2D] Frq Gen. / GN 50 / DR 54 / FA 8 / P 90



HERAW10

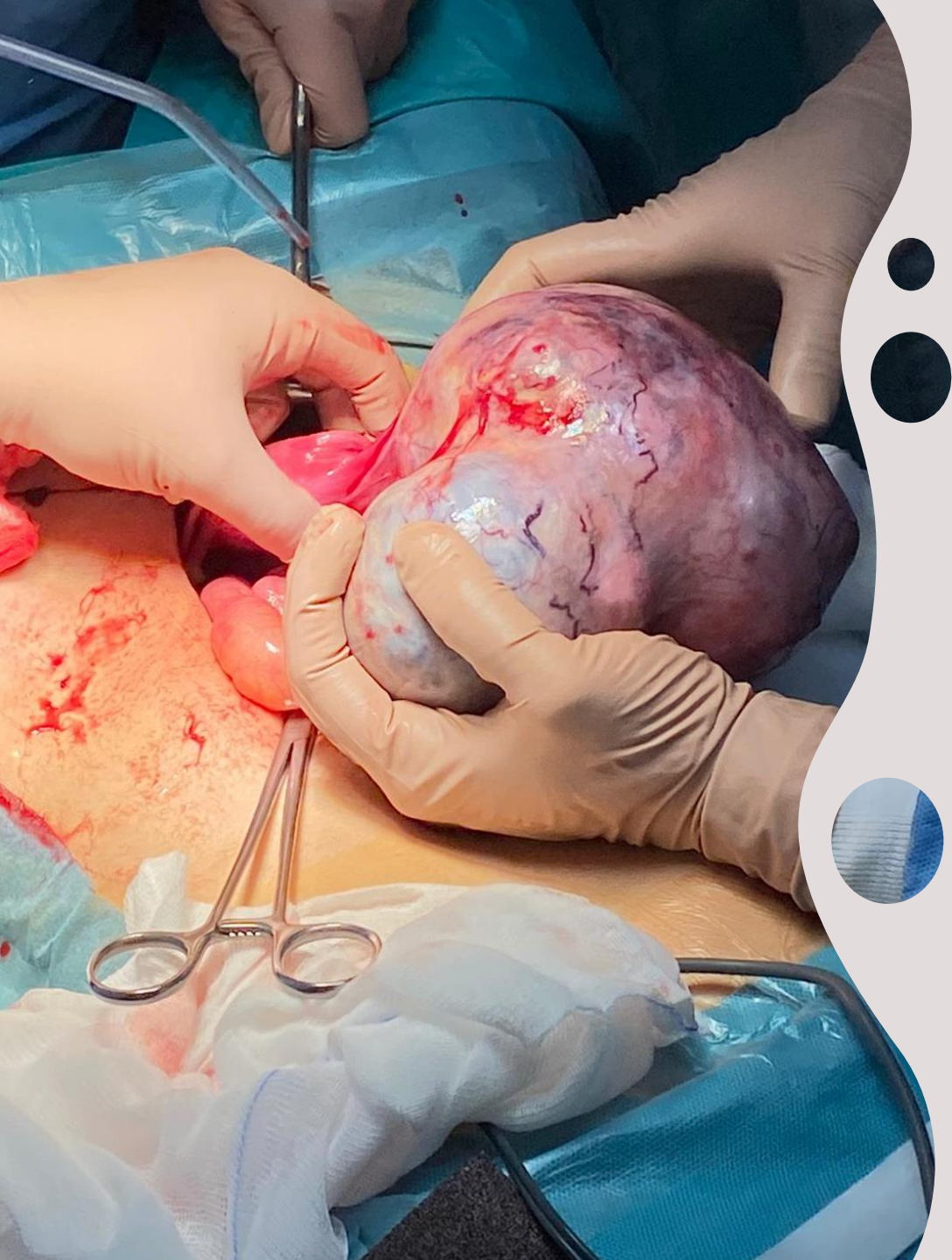


Gynecology / UTERO 1 / EV3-10B / FPS30 / 6.0cm / MI1.0 / TIs0.2 / 23-11-2020 11:17:05
[2D] Frq Gen. / GN 50 / DR 54 / FA 8 / P 90



HERAW10





TERATOMA IMMATURO

Teratoma contenente un quantitativo variabile di tessuto immaturo di tipo embrionale (neuroepitelio)



Neuroepiteliale:

Rosette
circondate da
cellule basofile
con mitosi

Mesenchimale:

Cartilagine,
grasso, osso,
muscolo striato
(immaturi)

Endoderma:

t. epatico, t.
intestinale, t.
renale (tipo
tumore di Wilms)

Grading

GRADO 1

Foci immaturi < 1 per campo a basso ingrandimento (40 x) in ogni vetrino

GRADO 2

Foci immaturi **1-3** per campo a basso ingrandimento (40 x) in ogni vetrino

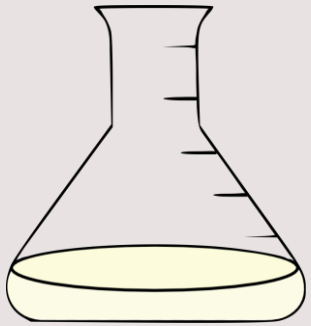
GRADO 3

Foci immaturi > 3 per campo a basso ingrandimento (40 x) in ogni vetrino



Bambine

**Giovani donne (<20
aa)**



AFP

**CA125 (rialzo
aspecifico)**



**Monolaterale
(12-15% bilaterale)
Grandi dimensioni
Rapida crescita
Addome acuto**



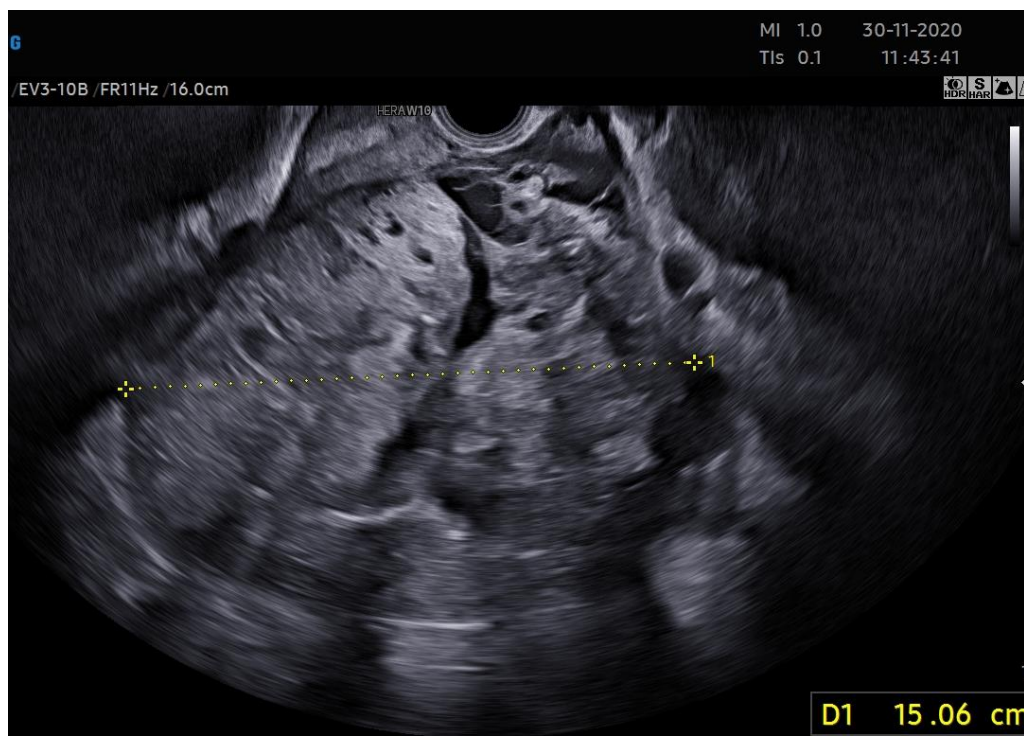
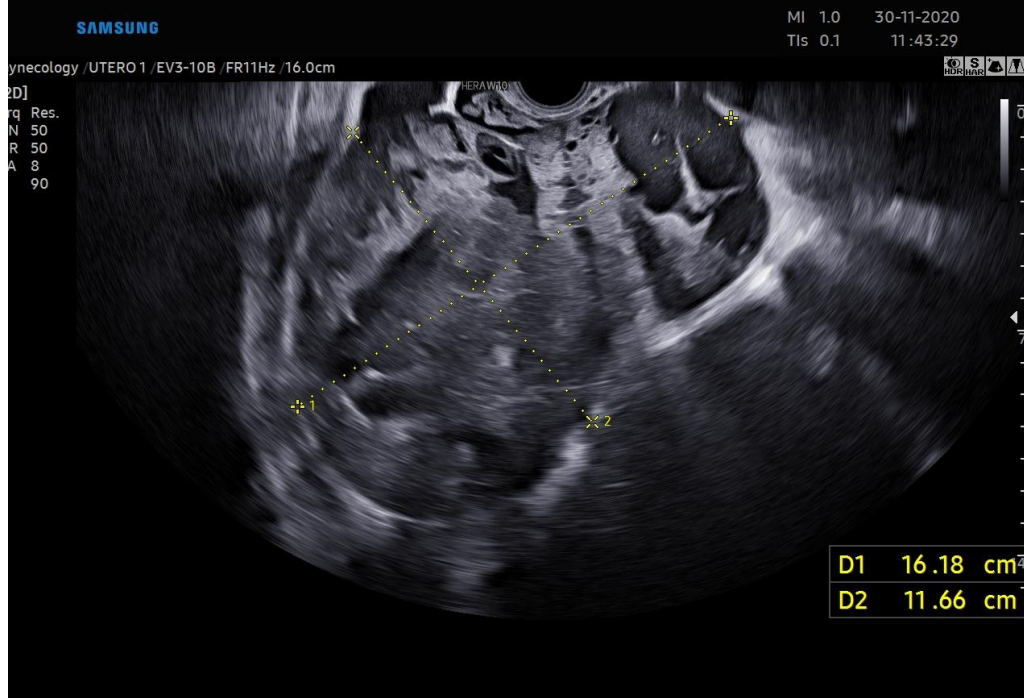
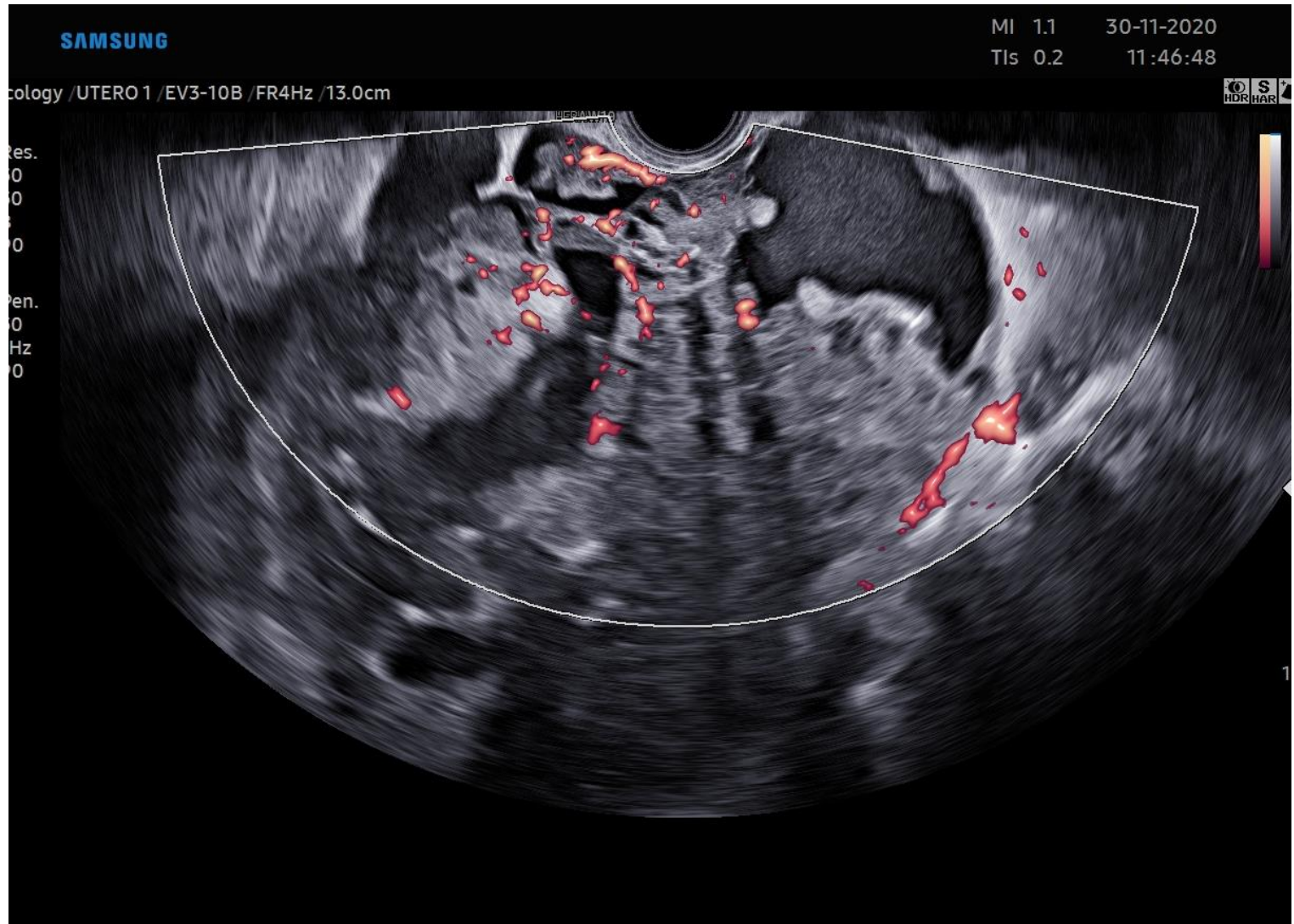
- **Massa SOLIDA o
MULTILOCULARE-
SOLIDA**
- **Ecogenicità bizzarra
della componente
solida**
- **SHADOWS irregolari**

HERAW10

17 aa

Autoriscontro di massa addominale palpabile

CA125 = 38; AFP = 17.652



Gynecology / UTERO1 / EV3-10B / FPS16 / 11.0cm / MI1.1 / TIs0.2 / 30-11-2020 11:42:37
[2D] Frq Res./GN 50/DR 50/FA 8/P 90



HERAW10

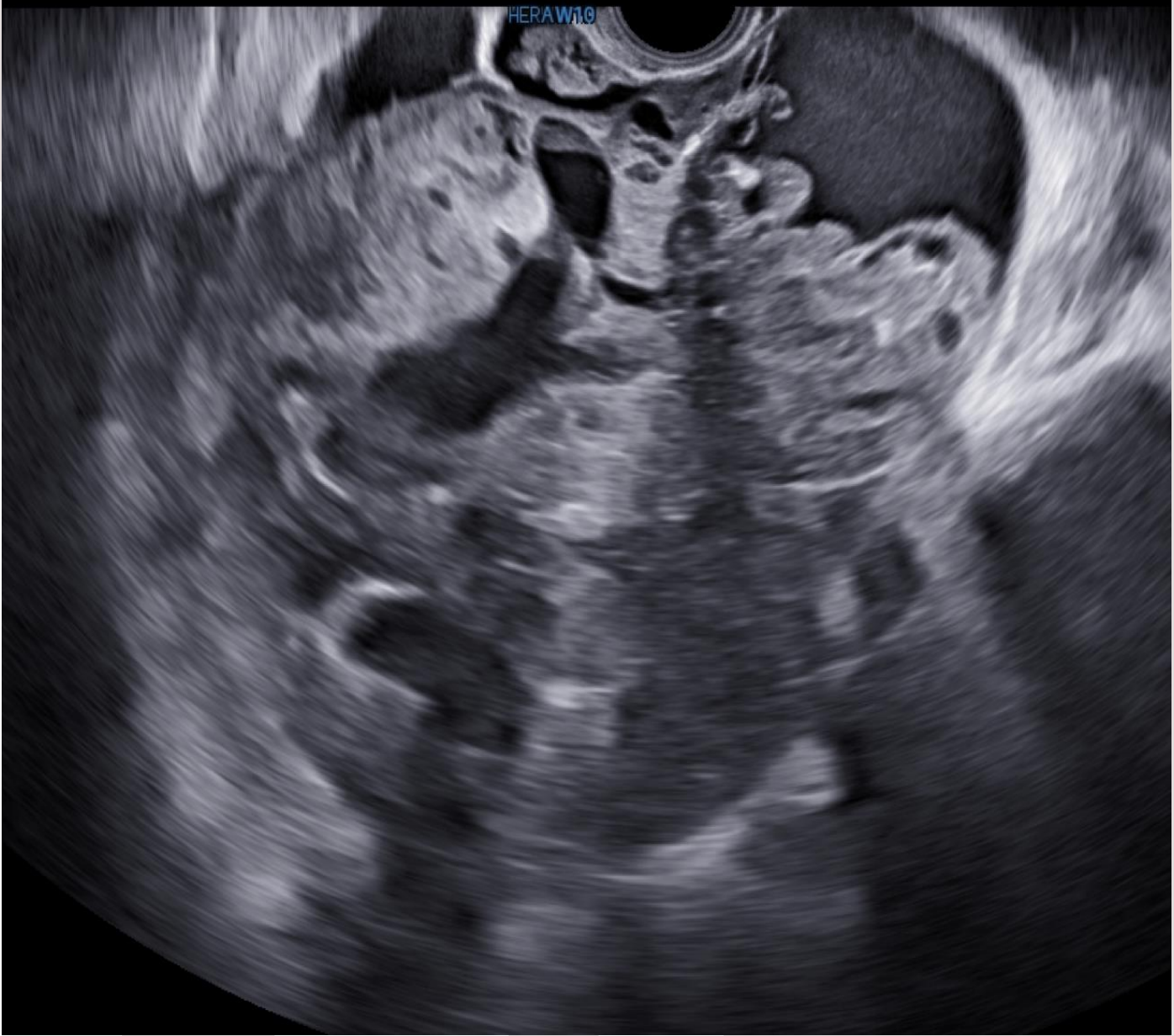


Gynecology / UTERO 1 / EV3-10B / FPS26 / 6.0cm / MI1.2 / TIs0.2 / 30-11-2020 11:42:17
[2D] Frq Res./GN 50/DR 50/FA 8/P 90



HERAW10





2D G50/DR126/FA6/P90/2.2MHz/Frq Gen./14.0cm



SAMSUNG
WS80A

40 aa

Dolore pelvico da 2 mesi

Riscontro di massa addomino-pelvica di 15 cm

CA125 = 549; AFP = 205

10

2D G42/DR104/FA10/P90/4.4MHz/Frq Gen./16.0cm



SAMSUNG
WS80A



7



14



2D G42/DR104/FA10/P90/4.1MHz/Frq Pen./16.0cm



SAMSUNG
WS80A



7

14

27 aa

Dolore pelvico e cicli irregolari

SAMSUNG

Gynecology /UTERO 1 /EV3-10B /FR6Hz /18.0cm

[2D]

Frq Pen.

GN 50

DR 50

FA 8

P 90

[PD]

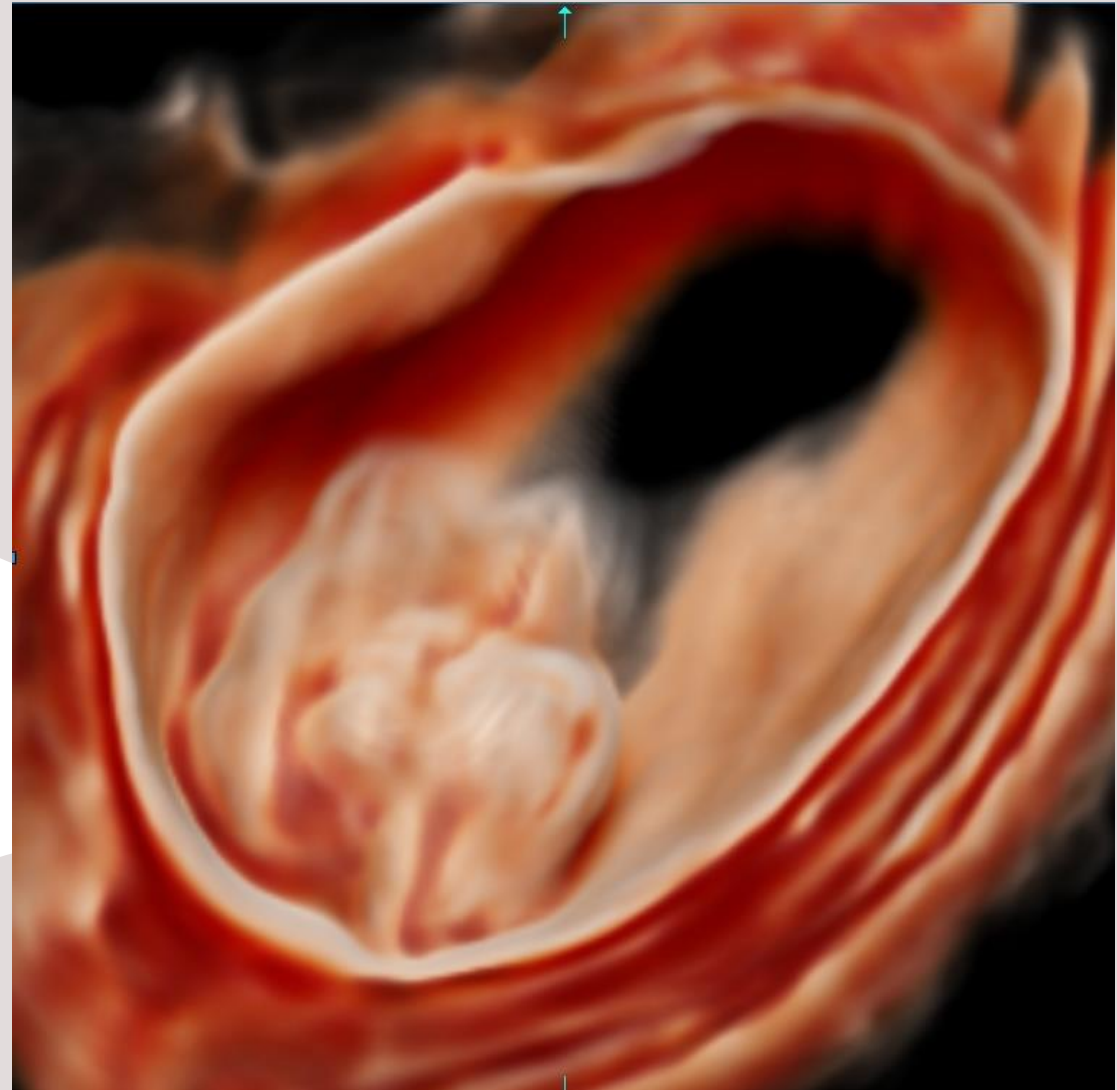
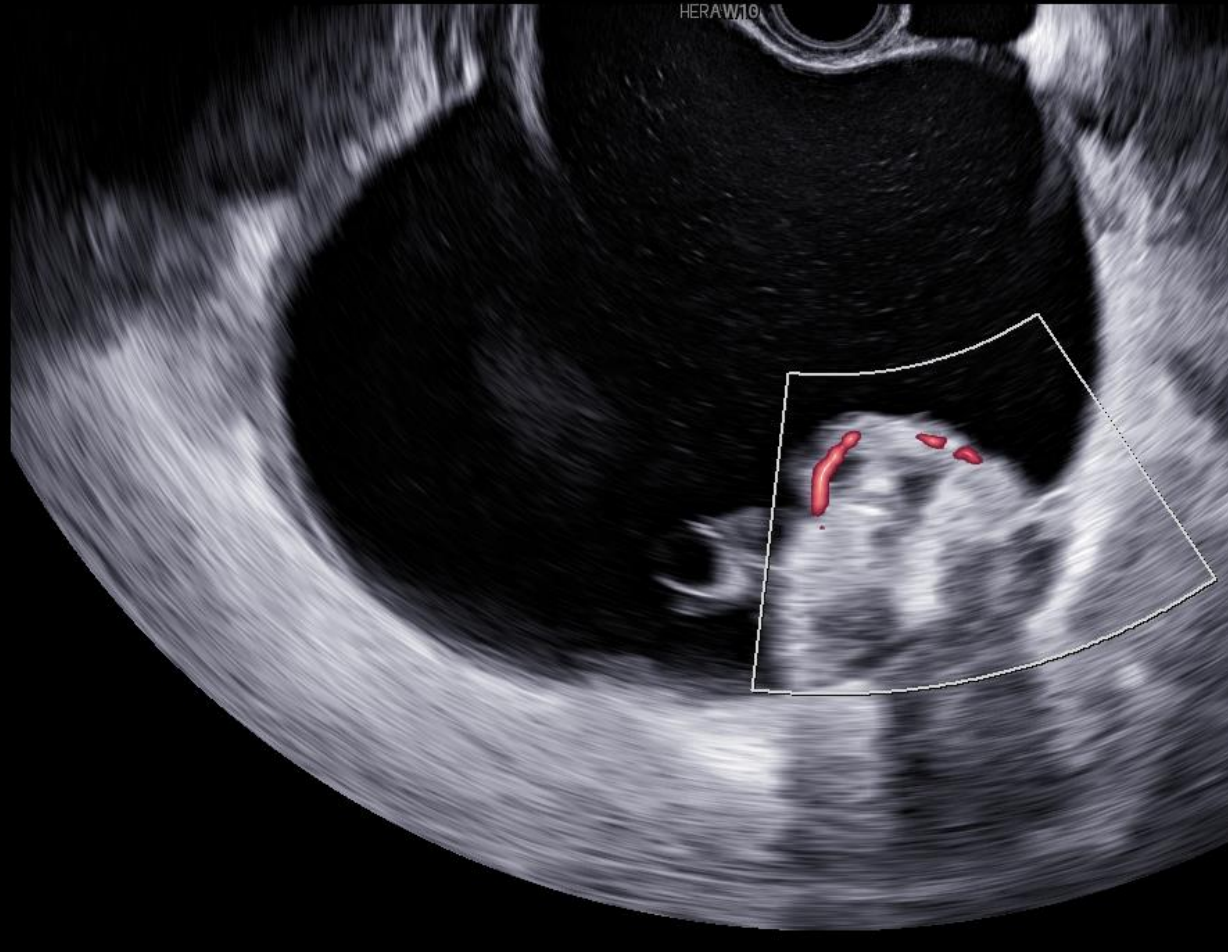
Frq Pen.

GN 50

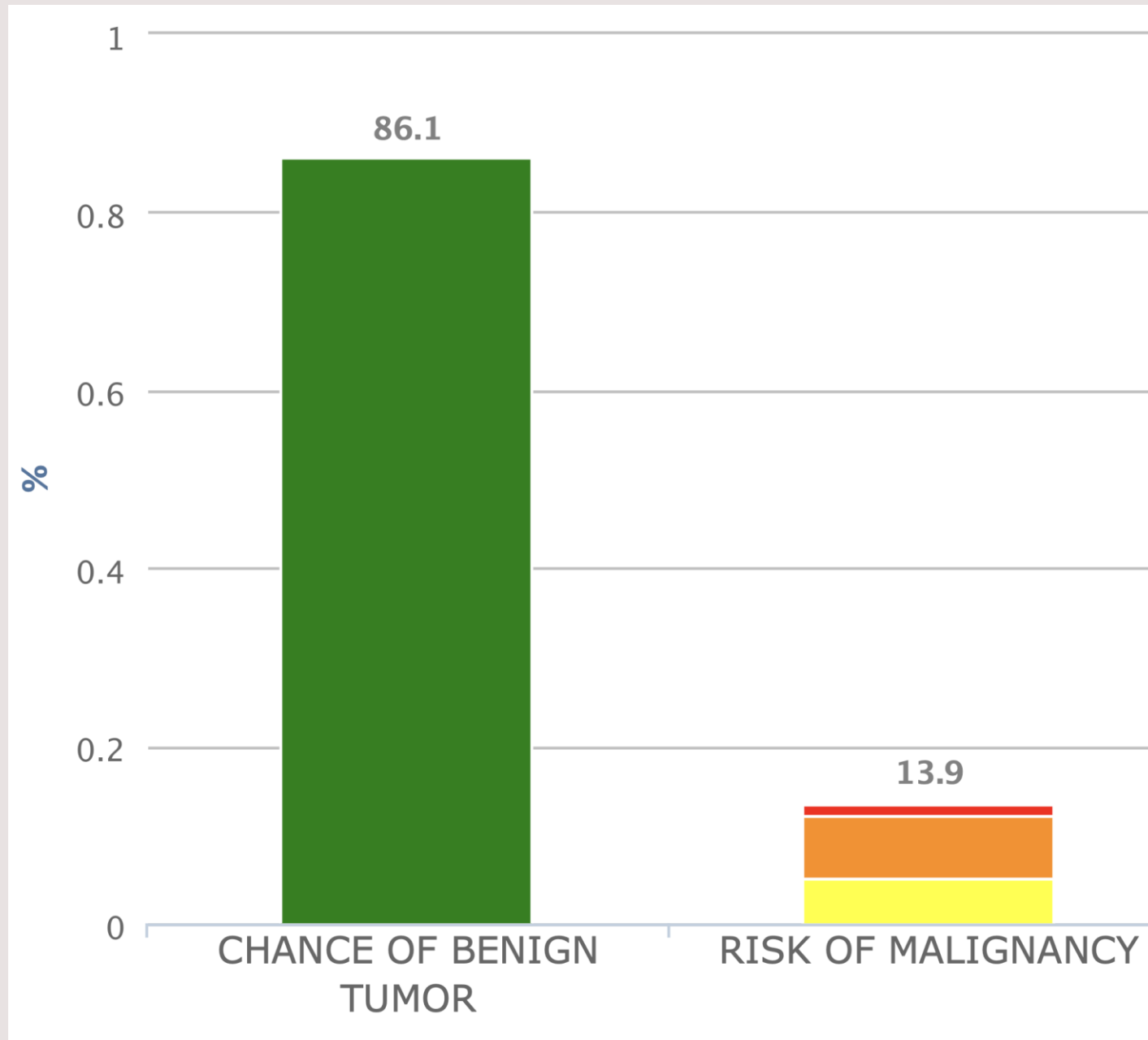
0.44kHz

P 90

HERAW10



- *Age: **27**
- *Oncological referral centre: **YES**
- * Max diameter of the lesion: **171**
- * Max diameter of the solid component: **64**
- *More than 10 locules: **NO**
- *Number of papillations: **ONE**
- *Acoustic shadows: **YES**
- *Ascites: **NO**
- *CA 125: **UNKNOWN**



GROWING TERAOMA SYNDROME

Growing Teratoma Syndrome and Gliomatosis Peritonei in a 15-Year-Old Girl With Immature Ovarian Teratoma: Case Report and Review of the Literature

Sawicka-Żukowska Małgorzata, MD PhD^{1,*}, Krętowska-Grunwald Anna, MD¹, Joanna Reszeć, MD², Maryna Krawczuk-Rybak, MD¹

What?

- (1) Ingrandimento o comparsa di masse retroperitoneali o in altre sedi durante o dopo la CHT
- (2) Normalizzazione markers
- (3) Presenza di cellule teratomatose mature nelle masse rimosse; un'ulteriore prosecuzione della CHT non ha effetto sulle masse

Why?

Teratoma immaturo

Chemioterapia

Teratoma maturo

Chemioterapia

Distrugge cellule maligne (benigne resistenti)

GLIOMATOSI PERITONEALE

Growing Teratoma Syndrome and Gliomatosis Peritonei in a 15-Year-Old Girl With Immature Ovarian Teratoma: Case Report and Review of the Literature

Sawicka-Żukowska Małgorzata, MD PhD^{1,*}, Krętowska-Grunwald Anna, MD¹, Joanna Reszeć, MD², Maryna Krawczuk-Rybak, MD¹

What?

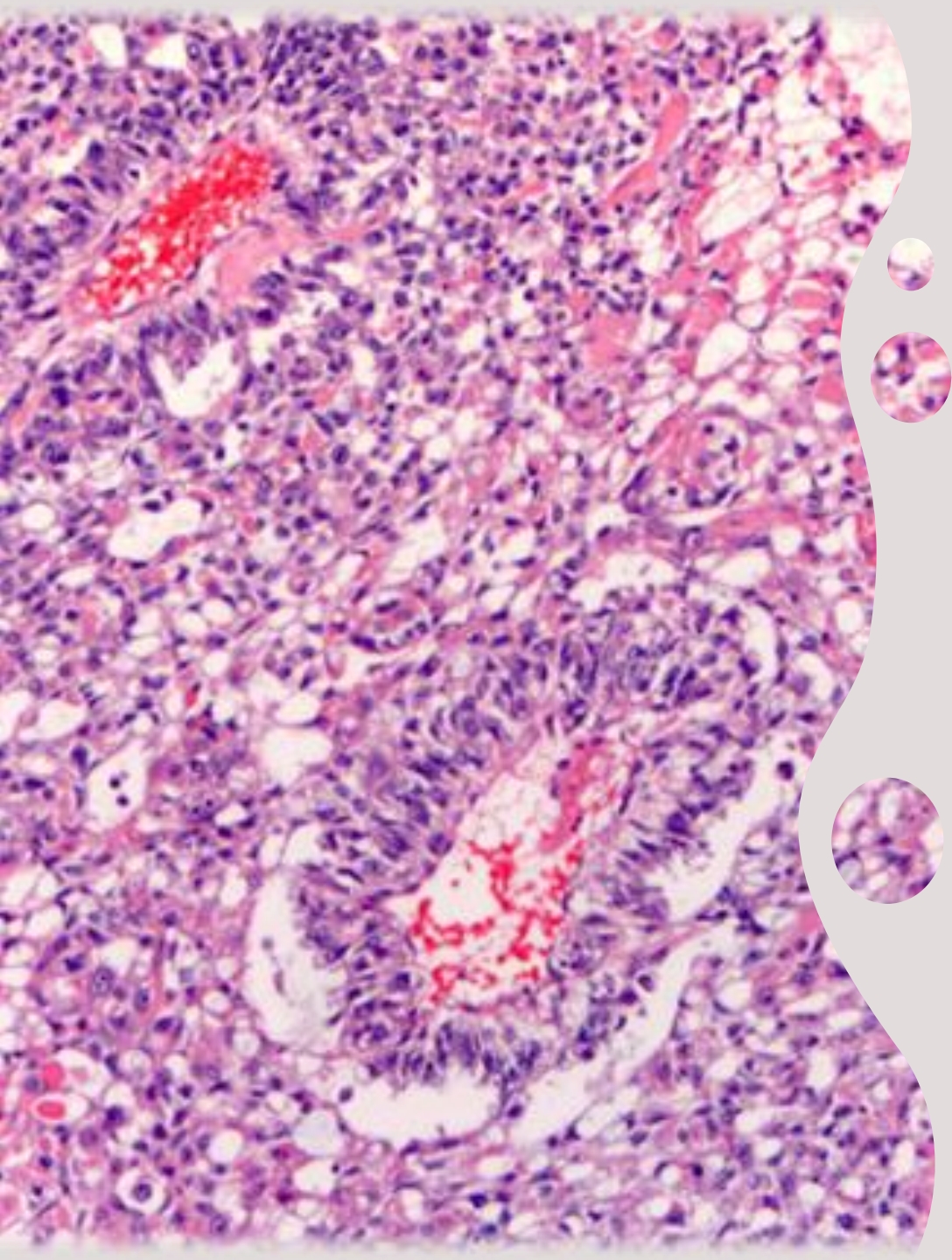
Presenza di impianti addominali di glia matura
Considerata come Teratoma grado 0

Why?

Rottura durante la prima chirurgia

Metastatizzazione per via linfatica o ematogena

Differenziazione di cellule peritoneali totipotenti in gliali indotta da secrezioni del teratoma



YOLK SAC TUMOR







Ultrasound Obstet Gynecol 2020; 56: 276–284

Published online in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/uog.22002.

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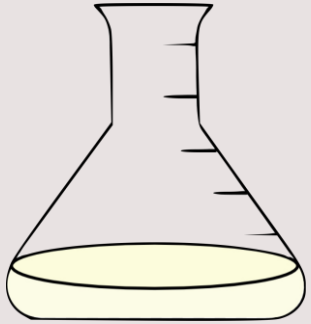


Imaging in gynecological disease (17): ultrasound features of malignant ovarian yolk sac tumors (endodermal sinus tumors)

P. ANFELTER^{1,2} , A. TESTA³, V. CHIAPPA⁴, W. FROYMAN⁵ , R. FRUSCIO⁶, S. GUERRIERO⁷ ,
J. L. ALCAZAR⁸ , F. MASCILLINI³, M. A. PASCUAL⁹ , M. SIBAL¹⁰, L. SAVELLI¹¹,
G. F. ZANNONI¹², D. TIMMERMAN⁵ and E. EPSTEIN^{1,2} 



Giovani (18-25 aa)
25% età prepuberale



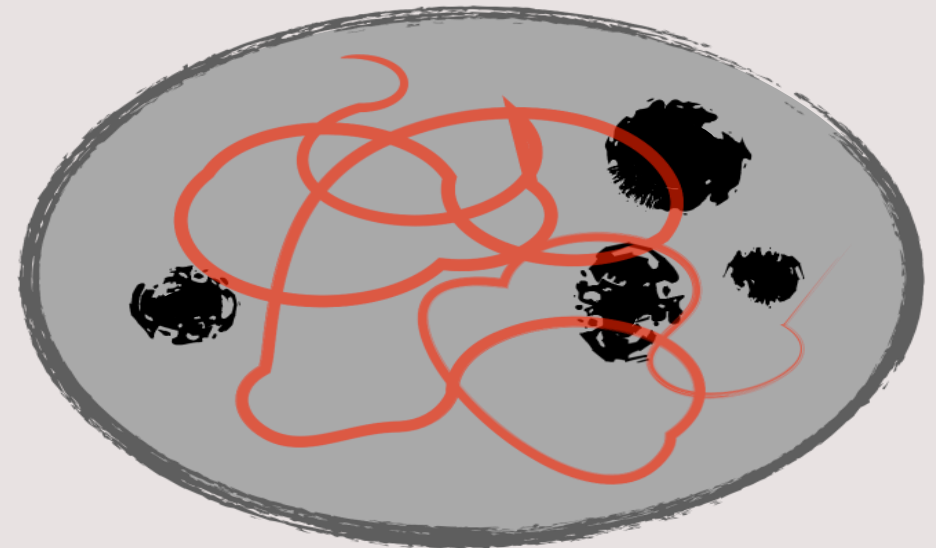
AFP
CA125 (rialzo
aspecifico)

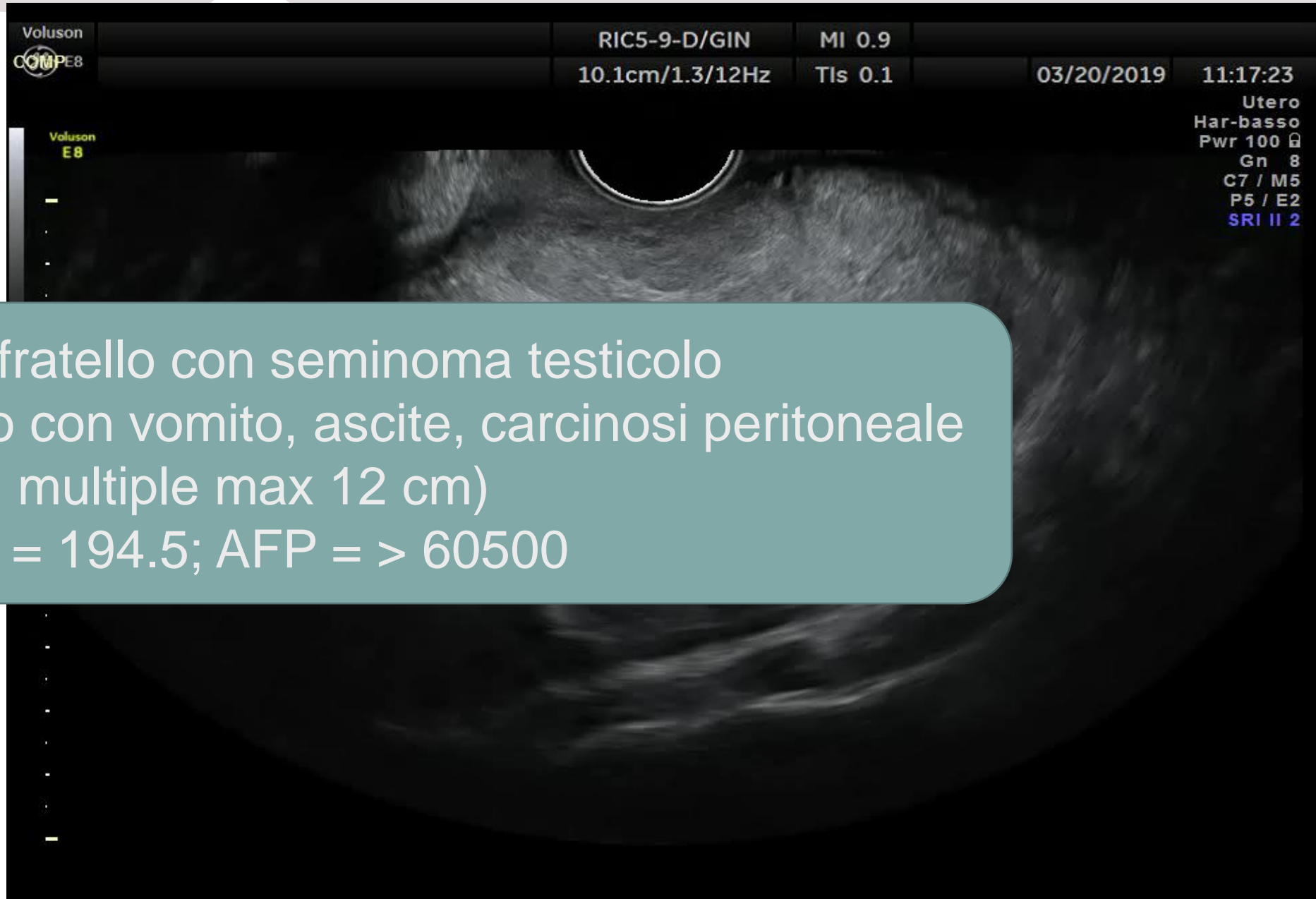


Monolaterale
Grandi dimensioni
Dolore
Sanguinamento
vaginale



- **Massa SOLIDA o**
MULTILOCALE-
SOLIDA
- **Color score 3-4**





39 aa, fratello con seminoma testicolo
Esordio con vomito, ascite, carcinosi peritoneale
(lesioni multiple max 12 cm)
CA125 = 194.5; AFP = > 60500

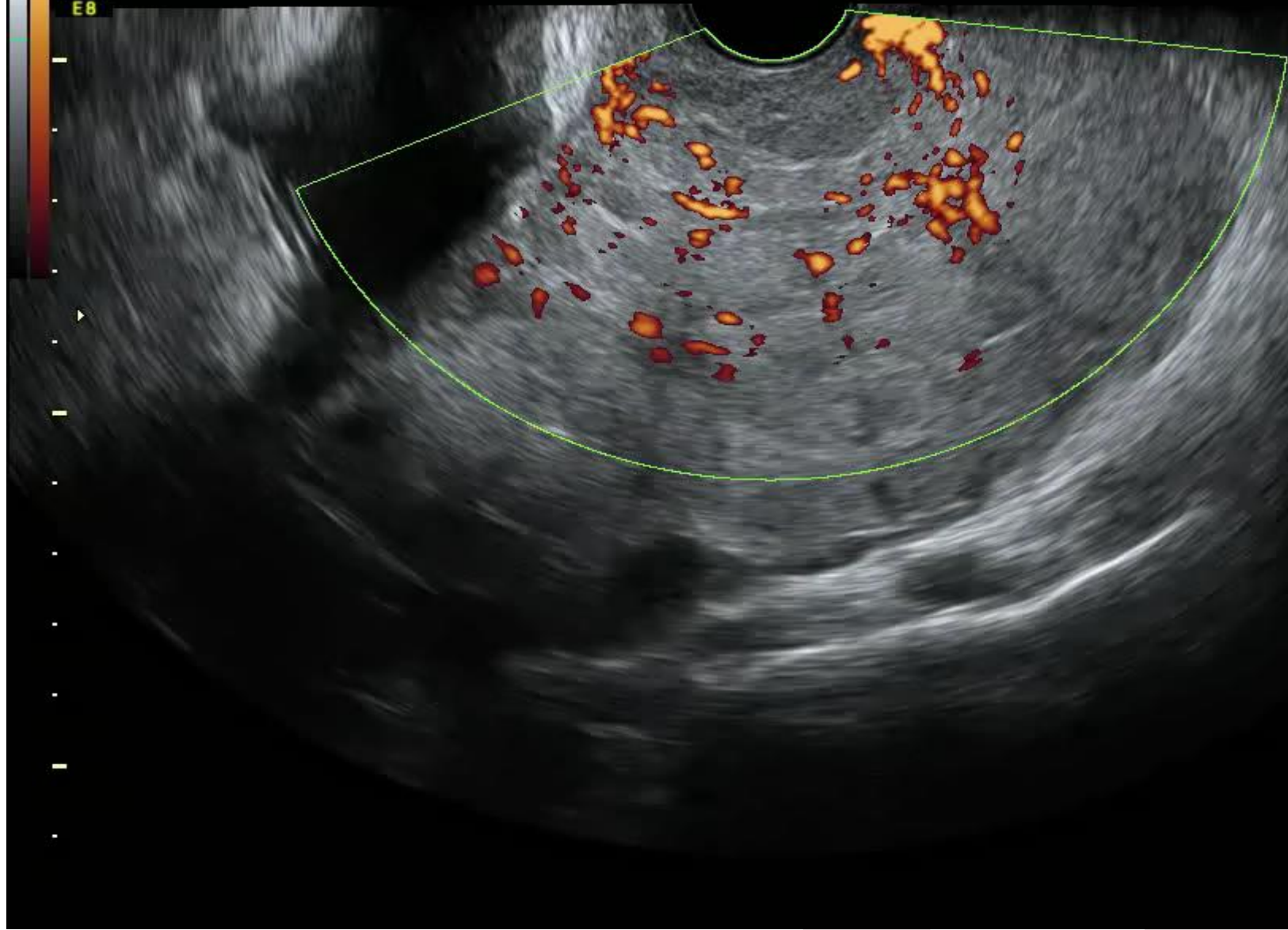
Voluson
COMP E8

RIC5-9-D/GIN
11.3cm/1.3/ 5Hz

MI 0.9
TIs 0.2

03/20/2019 11:18:20

Utero
10.00 - 4.00
Pwr 90 %
Gn 8
C7 / M5
P5 / E2
SRI II 2



Pwr 100 Ω
Gn -0.0
Frq med
Qual alto
WMF basso1
PRF 0.6kHz

Voluson

COMPE8

RIC5-9-D/GIN

MI 0.8

8.9cm/1.3/24Hz

TIs 0.1

03/20/2019

11:19:29

Utero
10.00 - 4.00

Pwr 100 dB

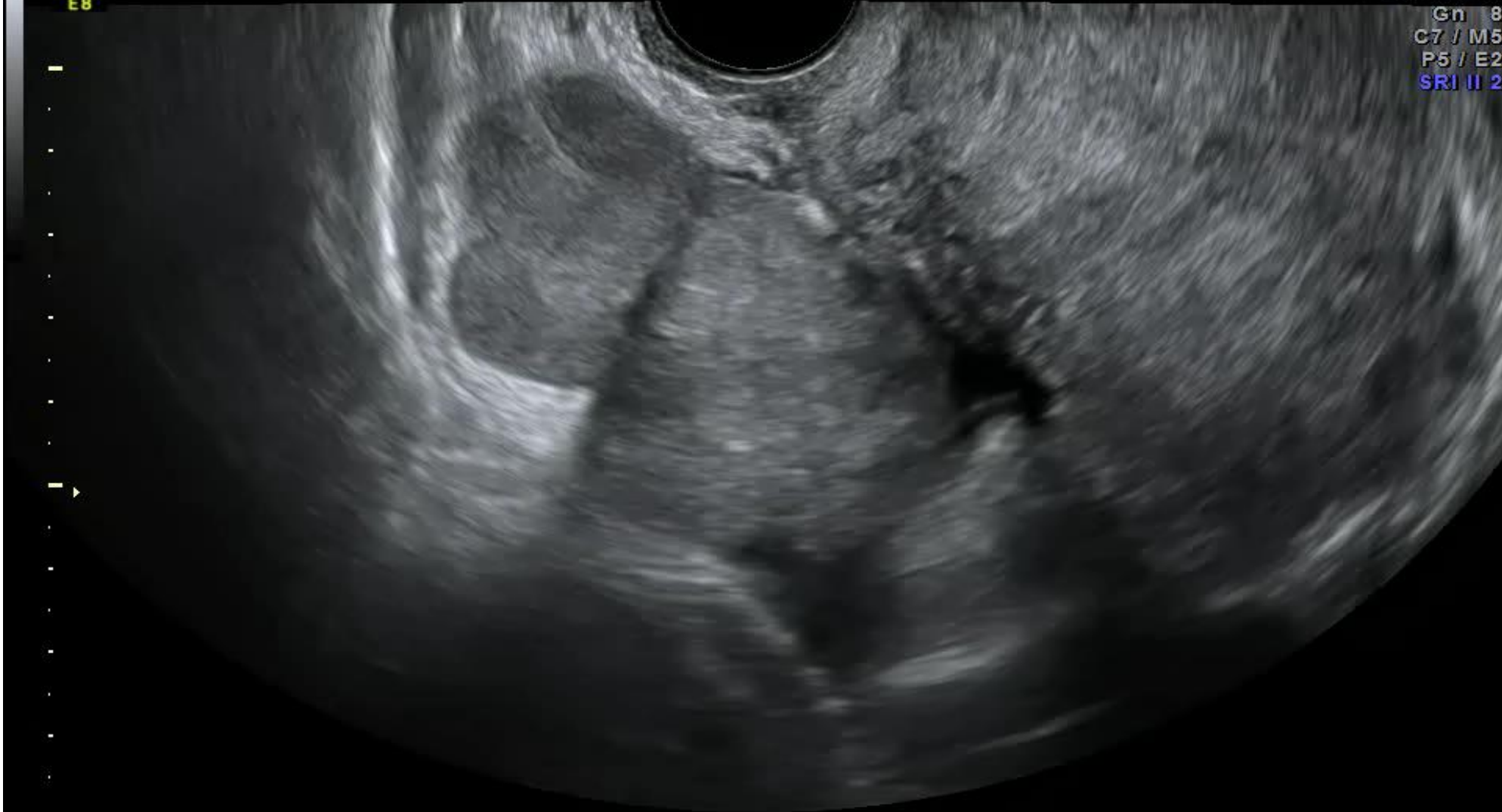
Gn 8

C7 / M5

P5 / E2

SRI II 2

Voluson
E8





Imaging in gynecological disease (17): ultrasound features of malignant ovarian yolk sac tumors (endodermal sinus tumors)

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21 pz

<i>Characteristic</i>	<i>Value</i>
Age (years)	25.0 (19.5–30.5)
Nulliparous	16 (76)
Personal history of ovarian cancer	1 (5)
FIGO Stage	
I	14 (67)
II	2 (10)
III	4 (19)
IV	1 (5)
Serum CA 125* (normal < 35 kU/L)	126 (35–227)
Serum AFP† (normal < 8 µg/L)	4755 (1071–25 303)



US features

Largest tumor diameter	157 mm
Max diameter solid component	110 mm
<i>Tumor type:</i>	
Solid	52%
Multilocular-solid	48%
Irregular lesion	90%
<i>Color score:</i>	
3	48%
4	48%
Shadowing	0
Hyperechoic solid tissue	89%

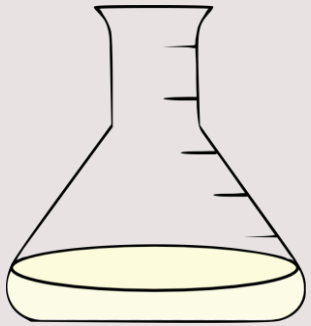


DISGERMINOMA

*Spesso riscontro incidentale
in corso di accertamenti per
infertilità (associazione con
disgenesia gonadica)*



**20-30 aa nel 75%
dei casi**



**LDH
CA125, PLAP, HCG**



**Monolaterale
Grandi dimensioni
Dolore pelvico
Incidental finding**



- **Massa SOLIDA
LOBULATA**
- **Superficie liscia**
- **Color score 3-4**





S

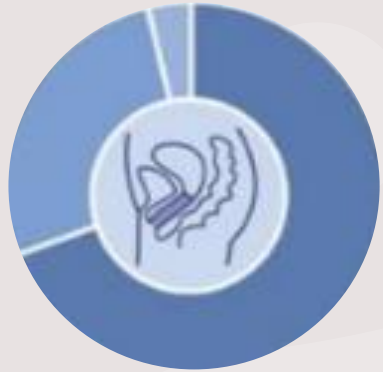


B RIS-A G --- CFM F 2.3 MHz G ---
TEI P 124mm XV +3 PRF 1.0 kHz
PRC 12/8/1 PRS 6+ PRC B/ 2 PRS 6
PST 0 C 2 FP 3

1
GENERALE

8
CA541





VAGINAL T.

- ✓ **84% 0-4 anni**
- ✓ **Rabdomiosarcom a embrionale**
- ✓ **10-year survival 86%**



CERVICAL T.

- ✓ **Istotipo più frequente rabdomiosarcom a embrionale seguito da adenocarcinoma**



UTERINE T.

- ✓ **Raro**
- ✓ **Gruppo eterogeneo di sarcomi, adenocarcinomi, choriocarcinomi**

MEDICAL ONCOLOGIST

PEDIATRI

C

PSYCHOLOGIST

GYN ONCOLOGIST

RADIOTHERAPIST

Grazie

RADIOLOGIST

TEACHER

PARENTS

FERTILITY PRESERVATION EXPERT

