PELVIC CONGESTION SYNDROME AND VULVAL VARICES – DIAGNOSIS, TREATMENT AND FOLLOW UP

ARINDAM BHARADWAZ

EMMA CORDSEN BUCHHAVE

PELVIC CONGESTION SYNDROME (PCS) AND VULVA-VARICES

- PCS is defined as chronic pelvic pain (CPP) caused by dilated and dysfunctional pelvic veins.
- Pelvic varicose veins were first described in the 1800s & the term PCS was First used in 1940s to describe chronic pelvic pain (CPP) due to vericose or ectatic pelvic veins.
- Vulvar varices (VV) are vericosities of the vulva, which may or may not be associated with vulvodynia, superficial dyspareunia and/or thromboembolic events.

-Jurga-Karwacka A, Karwacki GM, Schoetzau A, Zech CJ, Heinzelmann-Schwarz V, Schwab FD. A forgotten disease: Pelvic congestion syndrome as a cause of chronic lower abdominal pain. PLoS One. 2019 Apr 2;14(4):e0213834. doi: 10.1371/journal.pone.0213834. PMID: 30939134; PMCID: PMC6445463.

-Brown CL, Rizer M, Alexander R, Sharpe EE 3rd, Rochon PJ. Pelvic Congestion Syndrome: Systematic Review of Treatment Success. Semin Intervent Radiol. 2018 Mar;35(1):35-40. doi: 10.1055/s-0038-1636519. Epub 2018 Apr 5. PMID: 29628614; PMCID: PMC5886772.

- Saveliev VS, Pokrovsky AV, Zatyevakhin II, Kirienko AI. Rossiiskie klinicheskie rekomendatsii po diagnostike i lecheniyu khronicheskikh zabolevanii ven. Flebologia. 2013;7:18–20.

- Gavrilov SG. Vulvar varicosities: diagnosis, treatment, and prevention. Int J Womens Health. 2017 Jun 28;9:463-475. doi: 10.2147/IJWH.S126165. PMID: 28721102; PMCID: PMC5500487.

PCS – ORPHAN DISEASE

- No mention in Obs. & Gynec. Guidelines (RCOG - The Initial Management of Chronic Pelvic Pain - ***Green–top Guideline No. 41, May 2012***)
- Blue Cross- Blue Shield health Insurance Policy - Ovarian and Internal Iliac Vein Embolization is considered investigational as a treatment of Pelvic Congestion Syndrome. BCBSNC does not provide coverage for investigational services or procedures. - ***Last review March 2021***
- ***Systematic Review (Obstet Gynecol Surv. 2010)*** - recommended controlled trials between embolotherapy versus hormonal or other noninvasive treatments.
- The ***Society for Vascular Surgery*** and the ***American Venous Forum*** - Recommend embolization with coils, plugs or sclerotherapy alone or in combination (**Evidence Grade IIB**) – November 18, **2014**
- NHS and HTA (Health technology assessment) **2016** – RCT/systematic review to establish
  - Relationship between CPP & Radiological Evidence
  - Effectiveness of Embolization/Sclerotherapy

1. https://www.rcog.org.uk/globalassets/documents/guidelines/gtg_41.pdf
2. https://www.bluecrossnc.com/sites/default/files/document/attachment/services/public/pdfs/medicalpolicy/ovarian_and_internal_iliac_vein_embolization.pdf
3.Tu FF, Hahn D, Steege JF. Pelvic congestion syndrome-associated pelvic pain: a systematic review of diagnosis and management. Obstet Gynecol Surv. 2010 May;65(5):332-40. doi: 10.1097/OGX.0b013e3181e0976f. PMID: 20591203.
4.Gloviczki P et al. Society for Vascular Surgery; American Venous Forum. The care of patients with varicose veins and associated chronic venous diseases: clinical practice guidelines of the Society for Vascular Surgery and the American Venous Forum. J Vasc Surg. 2011 May;53(5 Suppl):2S-48S. doi: 10.1016/j.jvs.2011.01.079. PMID: 21536172.
5.Daniels JP, Champaneria R, Shah L, Gupta JK, Birch J, Moss JG. Effectiveness of Embolization or Sclerotherapy of Pelvic Veins for Reducing Chronic Pelvic Pain: A Systematic Review. J Vasc Interv Radiol. 2016 Oct;27(10):1478-1486.e8. doi: 10.1016/j.jvir.2016.04.016. Epub 2016 Jul 7. PMID: 27397619.

VULVAL VARICES

* Limited literature
* The incidence of VV is aproximately 4%
*But with PCS it rises to about 24% - 40%, particularly in premenopausal, multiparous women between 20 and 45 years
* The incidence rises to a whopping 72% at a mean parity of 6

Mechanisms

* Venous compression by a gravid uterus
* Hormonal venous dilatation
* Increased plasma volume and blood flow

Kim AS, Greyling LA, Davis LS. Vulvar Varicosities: A Review. Dermatol Surg. 2017 Mar;43(3):351-356. doi: 10.1097/DSS.0000000000001008. PMID: 28005626.

SYMPTOMATOLOGY

Pelvic congestion syndrome:

- Pain worsened by menstruation, pregnancy, prolonged standing and sitting
- Pain relief in supine position
- Dyspareunia
- Sensation of heaviness

Vulvar varicosities:

- Asymptomatic
- Pain
- Dyspareunia
- Bulging and pressure in the vulvar area
- Cosmetic issue

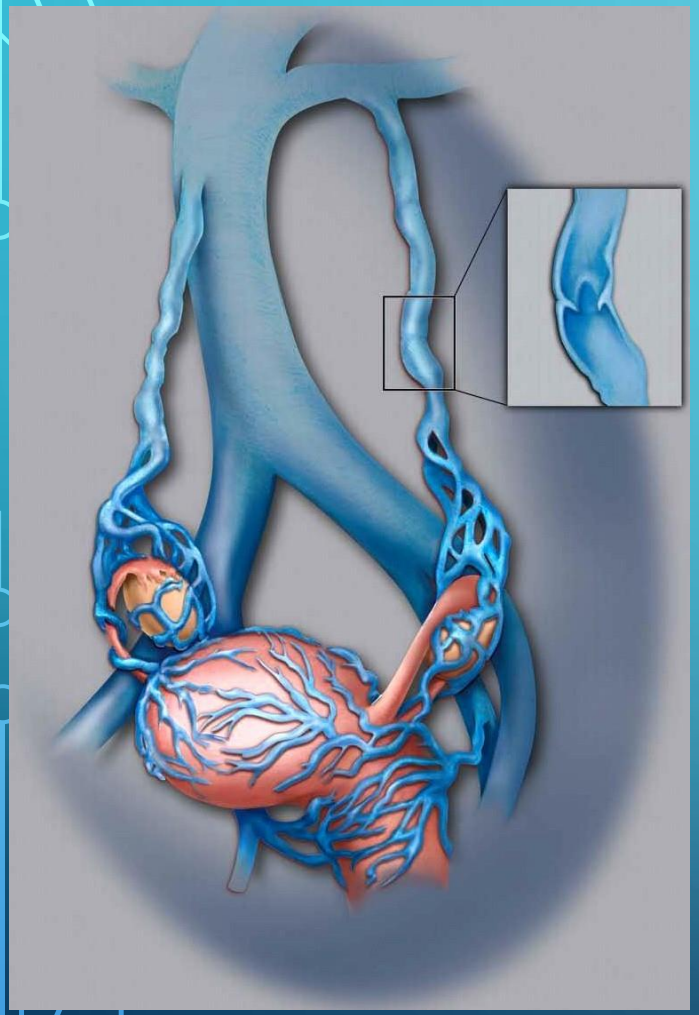
Bookwalter CA, VanBuren WM, Neisen MJ, Bjarnason H. *Imaging Appearance and Nonsurgical Management of Pelvic Venous Congestion Syndrome* . Radiographics 39(2): 596-608, 2019

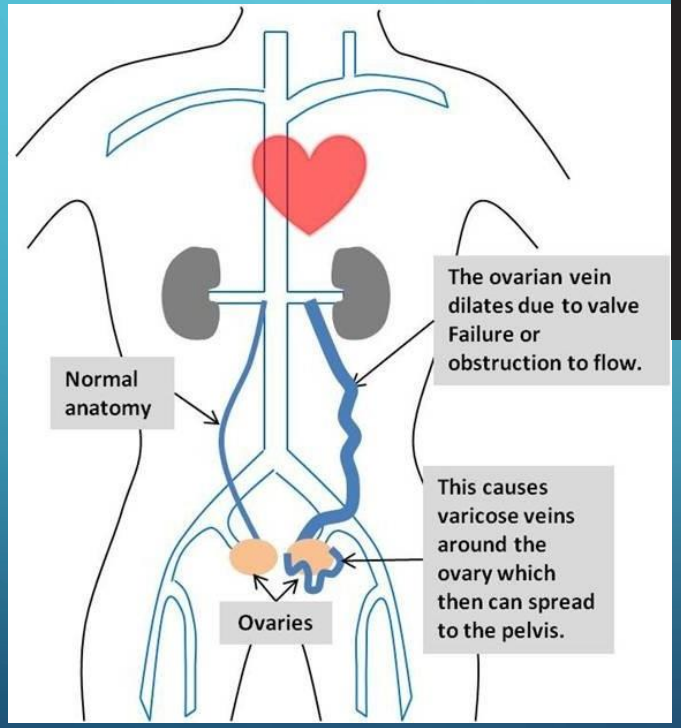
Borghi C, Dell'Atti L. *Pelvic congestion syndrome: the current state of the literature*. Arch Gynecol Obstet 293(2): 291-301, 2016

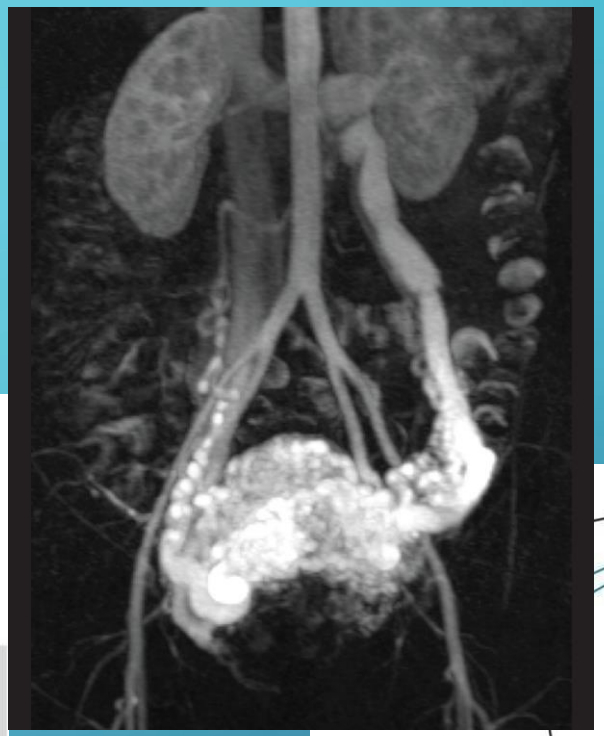
Kim AS, Greyling LA, Davis LS. *Vulvar Varicosities: A Review*. Dermatol Surg 43(3): 351-6, 2017

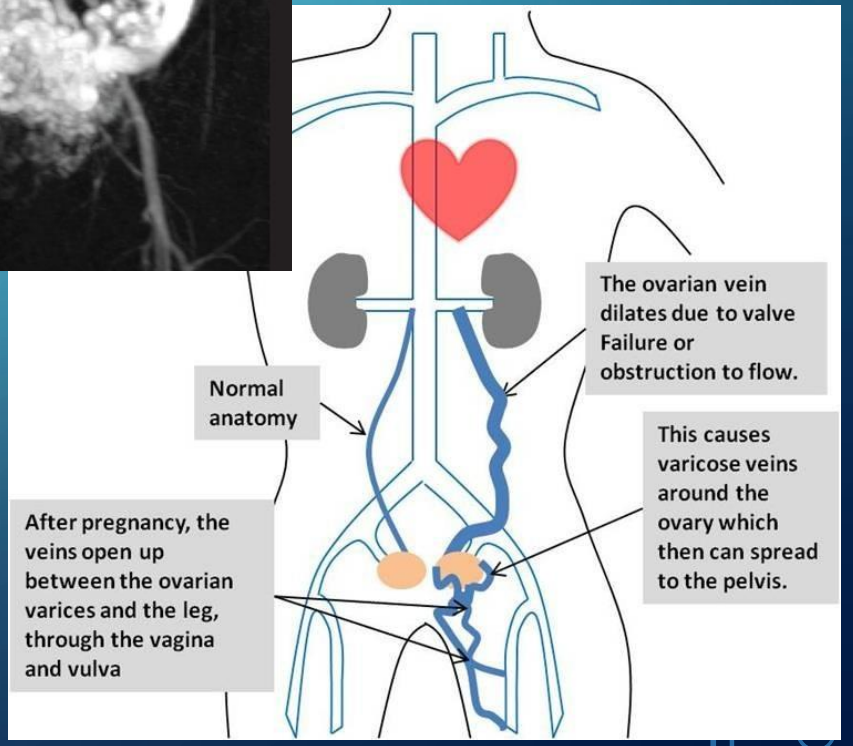
Al Wahbi AM. *Isolated large vulvar varicose veins in a non-pregnant woman*. SAGE Open Med Case Rep 4: 2050313x16672103, 2016

ANATOMY OF PELVIC CONGESTION SYNDROME (PCS)



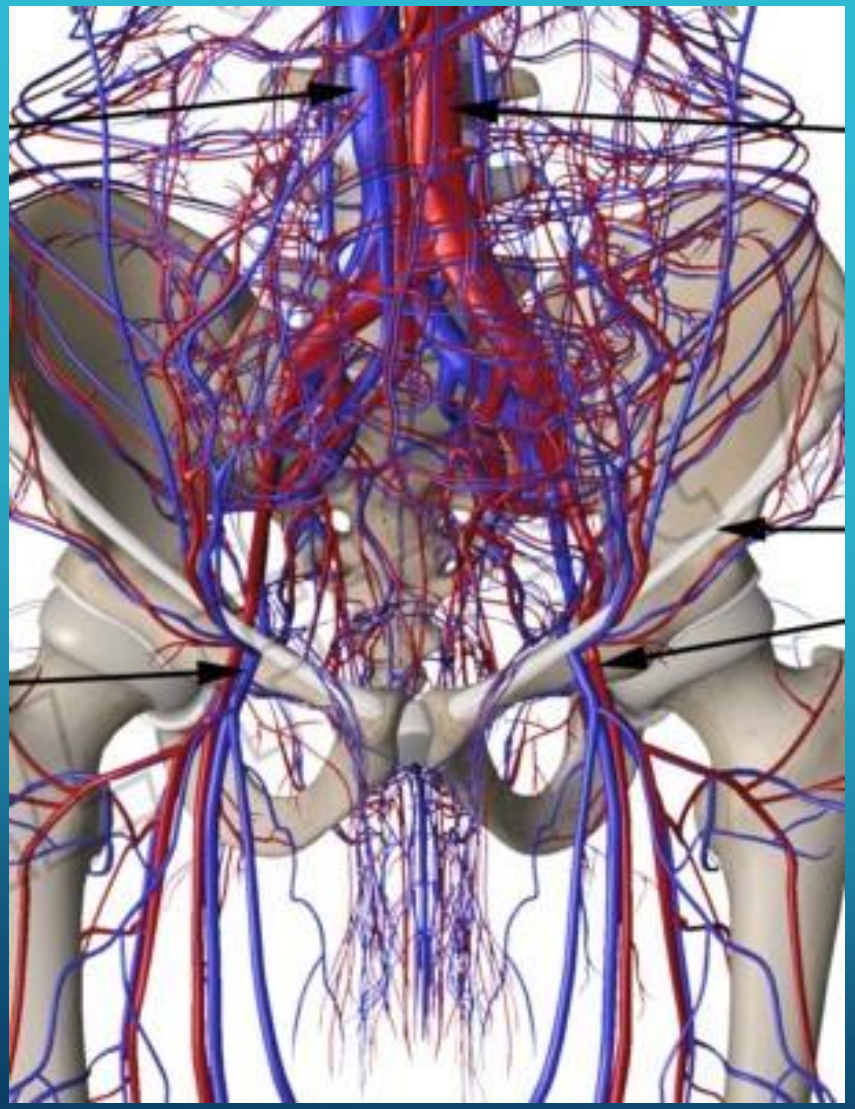


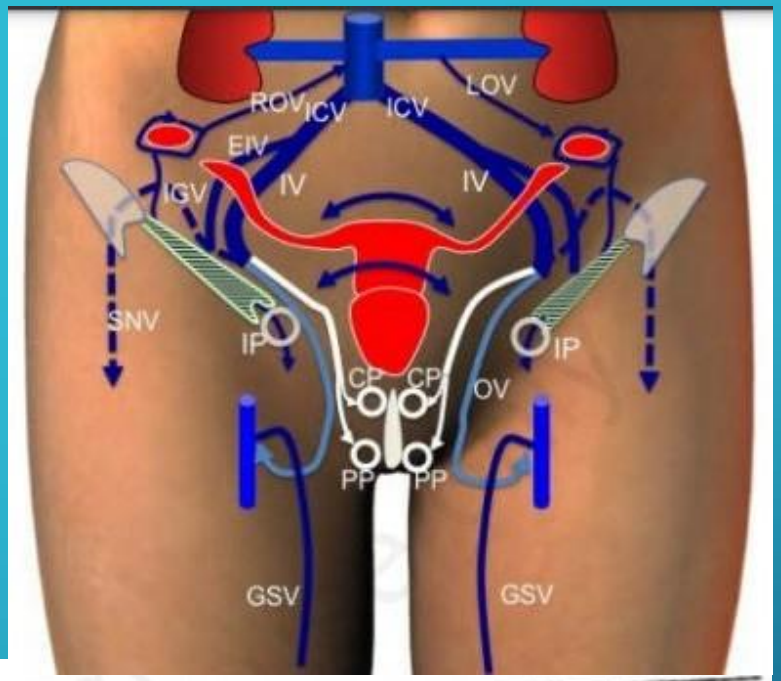


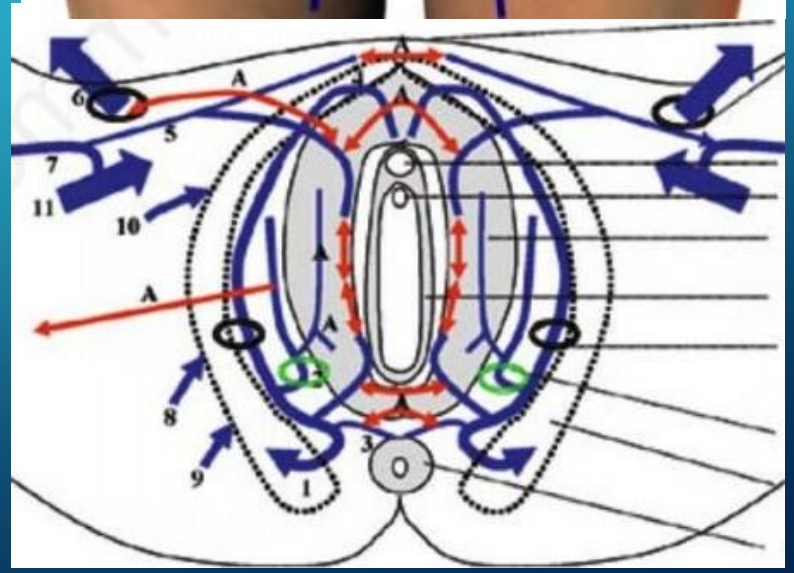


http://www.bsir.org/patients/pelvic-venous-congestion-syndrome/

ANATOMY OF VULVAL VARICES (VV)







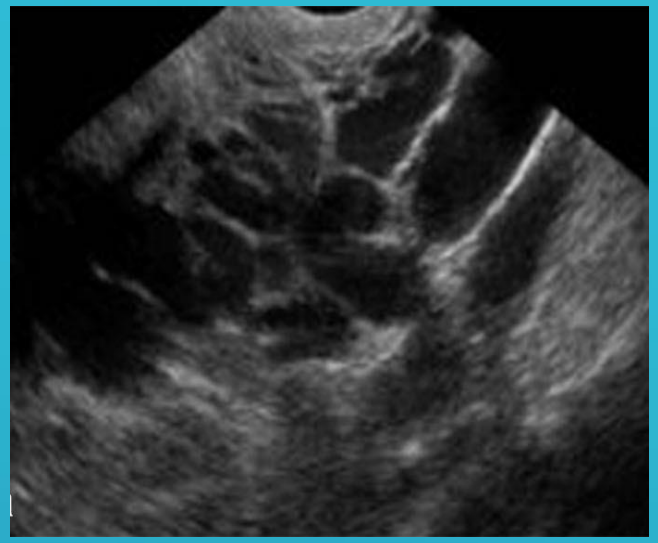
https://www.realfirstaid.co.uk/pelvis

Minimally-invasive procedure for pelvic leak points in women. Roberto Delfrate, Massimo Bricchi, Claude FranceschiVeins and Lymphatics 2019; 8:7789]

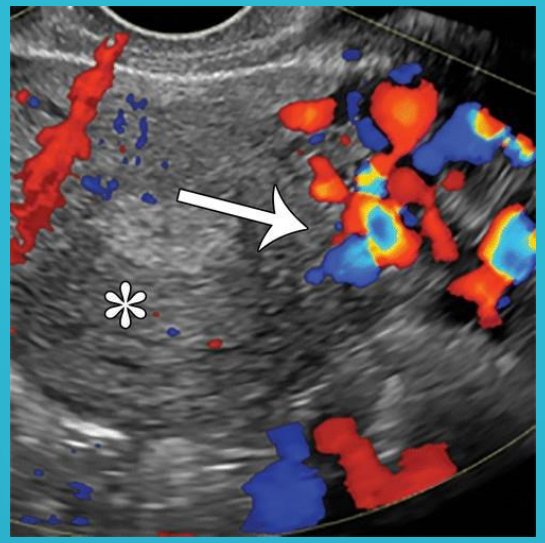
IMAGING EVALUATION OF PCS & VULVAL VARICES

USG	***Initial investigation,*** Rule out other pelvic pathology, inexepensive, easily available, scanning in upright position possible, ***dynamic*** real- time, measure ***direction*** and ***velocity*** of flow
MRI	Exclude other pathology, ***overview*** possible, dynamic evaluation possible, expensive, difficult to interpret
CT	Exclude other pathology, ***overview*** possible, Invaluable for ***treatment planning*** exclude anatomical variations.
Phlebography	***Golden standard***, diagnostic and therapeutic.
Laparoscopy	***Unrealiable***

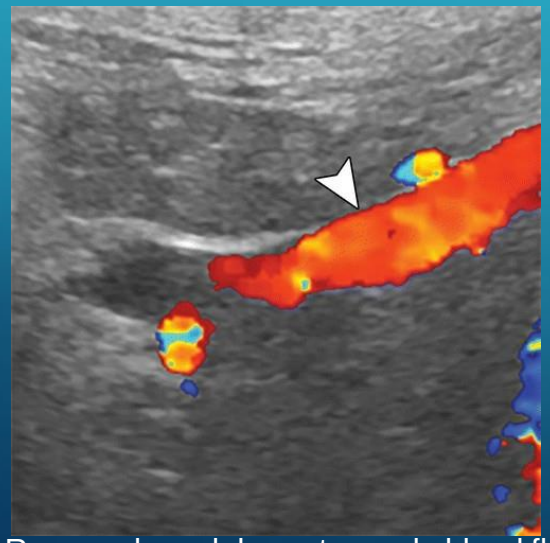
ULTRASOUND CRITERIA FOR PCS



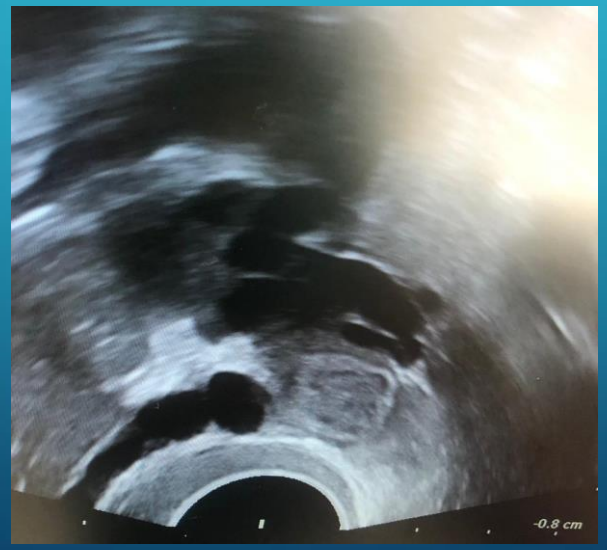
Presence of tortuous and dilated pelvic venous plexuses
Dilation of the left ovarian vein (more than 4-8 mm in diameter)



Variable duplex waveform in the varicoceles during the Valsalva's maneuver (implying valve incompetence)



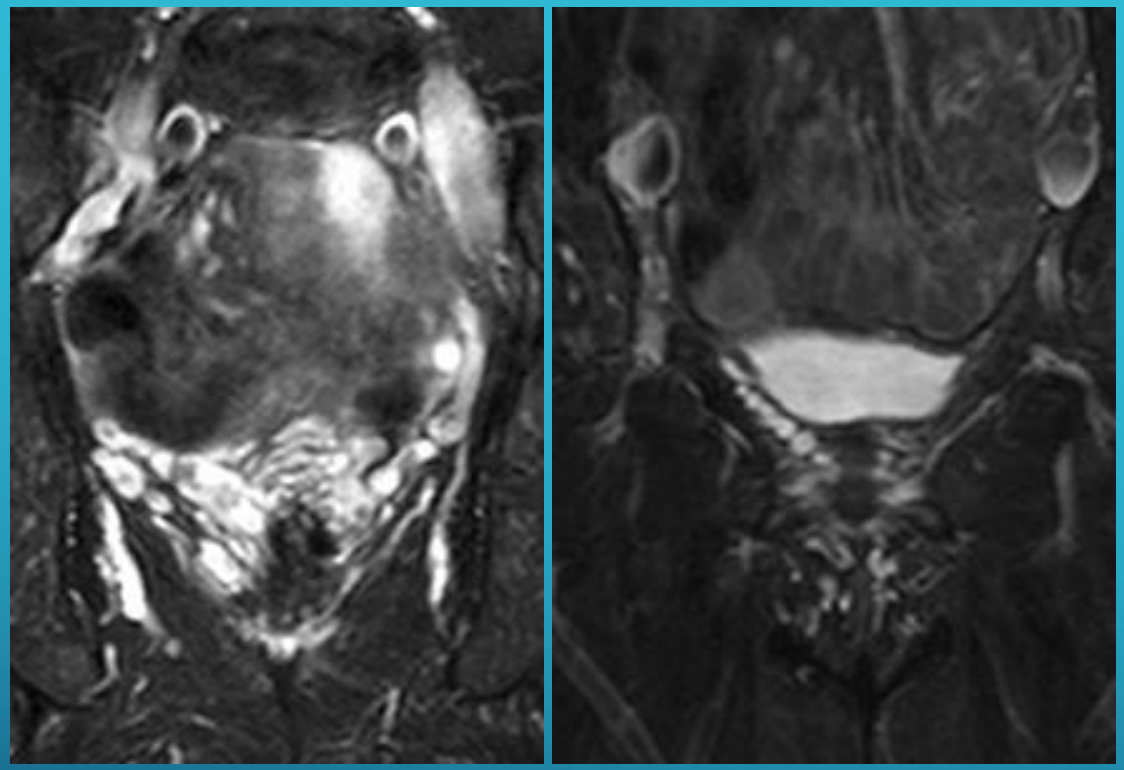
Reversed caudal or retrograde blood flow

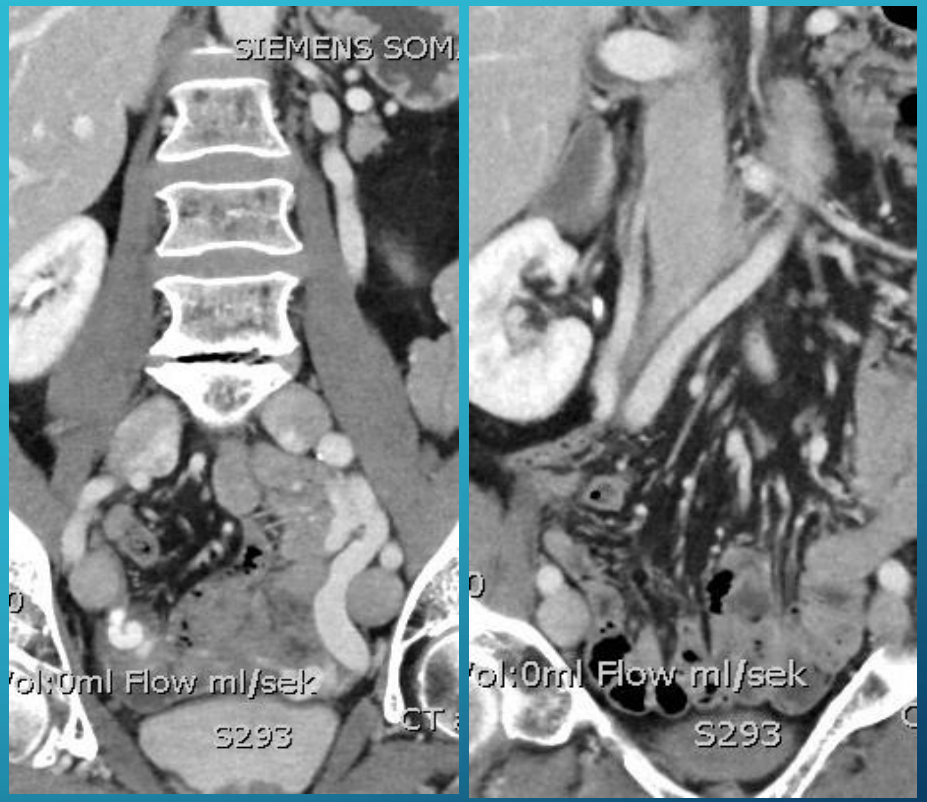


- Rule out uterine, ovarian or pelvic patholgy
- Dilation of the left ovarian vein (more than 4-8 mm in diameter)
- Reversed caudal or retrograde blood flow
- Slow blood flow (less than 3 cm/s)
- Presence of tortuous and dilated pelvic venous plexuses
- Dilated arcuate veins crossing the uterine myometrium that communicate with bilateral pelvic varicose veins
- Variable duplex waveform in the varicoceles during the Valsalva's maneuver (implying valve incompetence)
- Polycystic changes of the ovary, different from those of women with classic polycystic ovarian syndrome. The significance of this finding remains unclear.

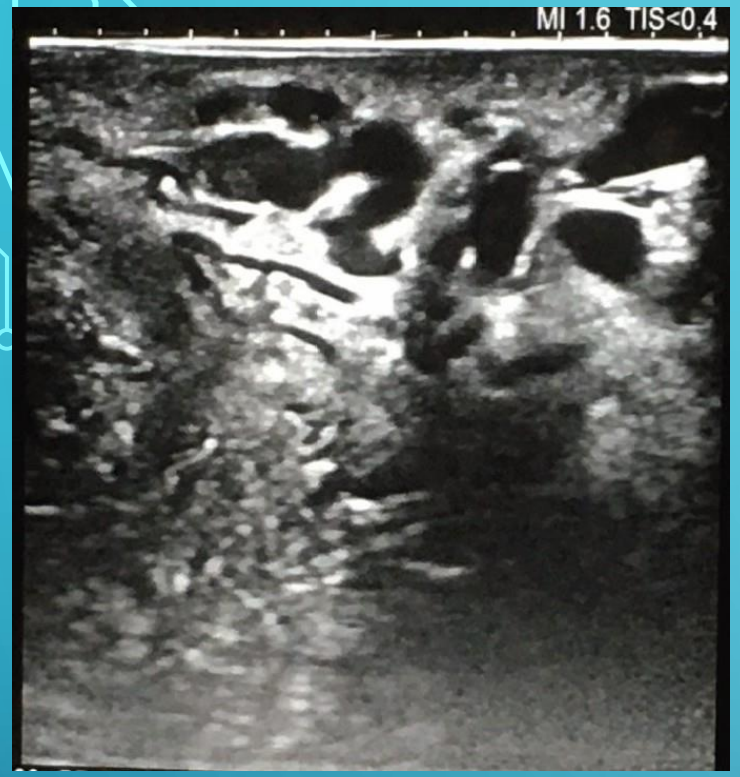
- Borghi C, Dell'Atti L. Pelvic congestion syndrome: the current state of the literature. Arch Gynecol Obstet. 2016 Feb;293(2):291-301. doi: 10.1007/s00404-015-3895-7. Epub 2015 Sep 24. PMID: 26404449.
- Bookwalter CA, VanBuren WM, Neisen MJ, Bjarnason H. Imaging Appearance and Nonsurgical Management of Pelvic Venous Congestion Syndrome. Radiographics. 2019 Mar-Apr;39(2):596-608. doi: 10.1148/rg.2019180159. PMID: 30844351.

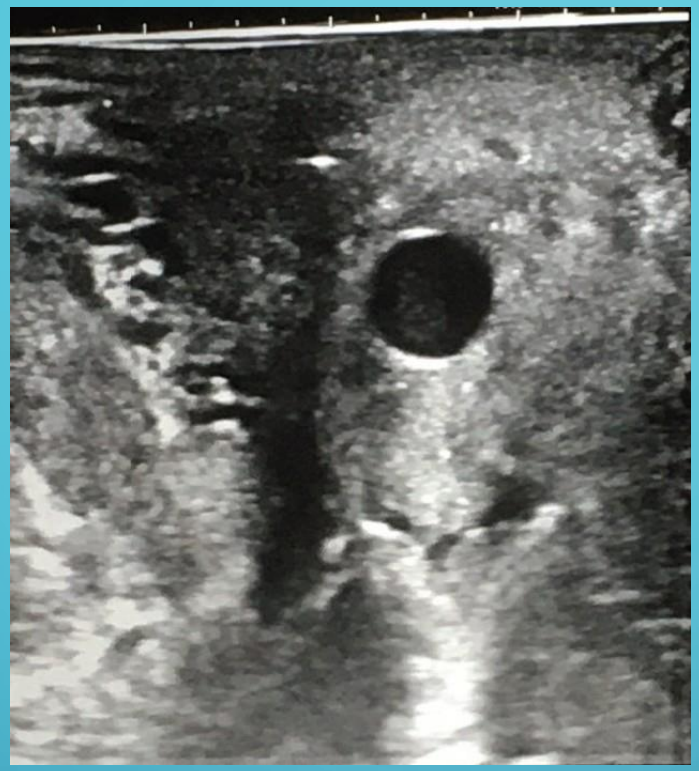
PCS – DIAGNOSTIC MR/CT/USG

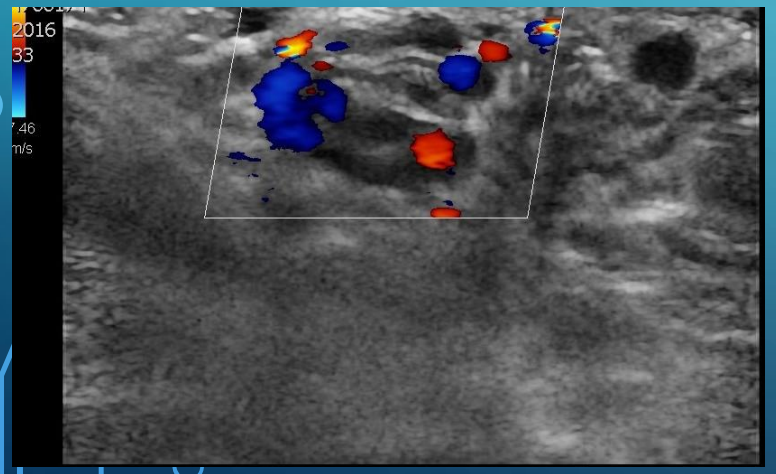




- Rule out uterine, ovarian or pelvic patholgy
- Dilation of the left ovarian vein (more than 4-8 mm in diameter)
- Presence of tortuous and dilated pelvic venous plexuses
- Dilated arcuate veins crossing the uterine myometrium that communicate with bilateral pelvic varicose veins, communication with varices in vulva, thigh. perineum etc.

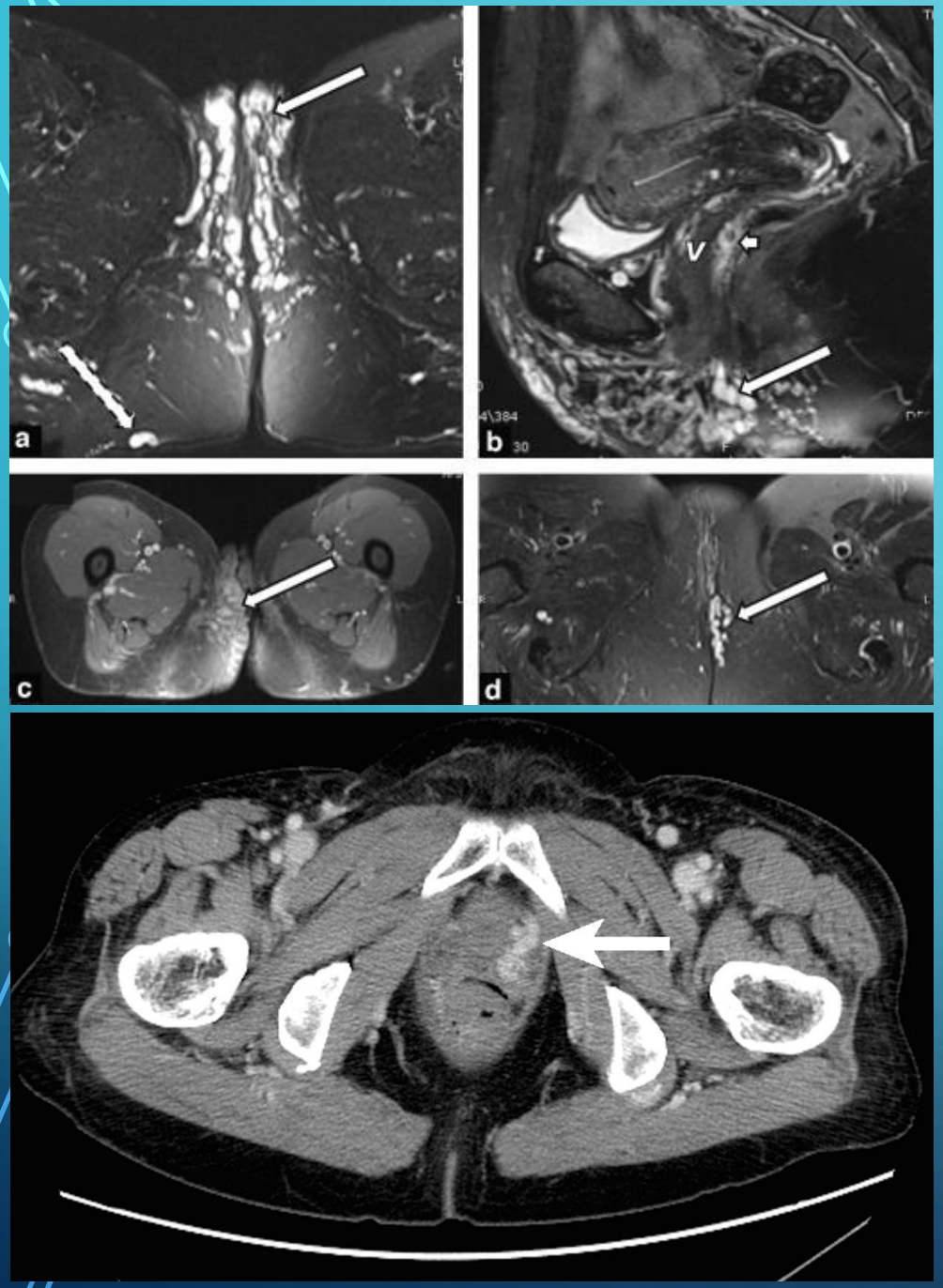






ULTRASOUND CRITERIA FOR VV

- Rule out uterine, ovarian or pelvic patholgy
- Dilated tortuous vulval veins
- Slow blood flow (less than 3 cm/s)
- Presence of tortuous and dilated pelvic venous plexuses and other criteria for PCS - if pt. has concomittant PCS
- Presence of SFJ (Sapheno-phemoral junction) incompetence in patients with LSV (long saphenous vein) varices.



CT & MR FOR VV

- CT or MR are not essential/must for isolated VV
- Adjunct diagnostic modalities
- Rule out uterine, ovarian or pelvic patholgy
- Presence of tortuous and dilated pelvic venous spaces in vulva
- Presence of tortuous and dilated pelvic venous plexuses and other criteria for PCS - if pt. has concomittant PCS
- Presence of other varicosities e.g. leg and perineal vericosities etc.

Leiber LM, Thouveny F, Bouvier A, Labriffe M, Berthier E, Aubé C, Willoteaux S. MRI and venographic aspects of pelvic venous insufficiency. Diagn Interv Imaging. 2014 Nov;95(11):1091-102. doi: 10.1016/j.diii.2014.01.012. Epub 2014 Mar 12. PMID: 24630150.

Kondo T, Uehara T, Noda K, Ohira Y, Ikusaka M. Vulvar varicosity. BMJ 2017;356:i6332 doi: https://doi.org/10.1136/bmj.i6332 (Published 26 January 2017)

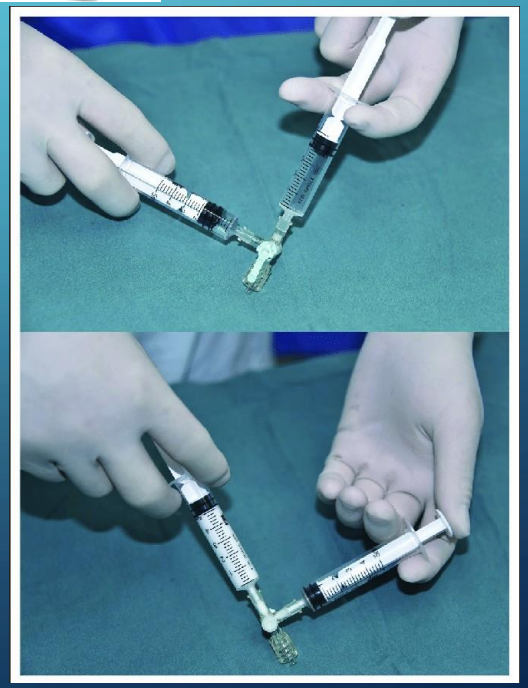
EMBOLIZATION OF PCS AND SCLEROTHERAPY FOR VV

PCS Performed under Local Anesthesia
Entry - Femoral or Arm
Left ovarian vein aproached via left renal vein
Right ovarian vein directly from IVC

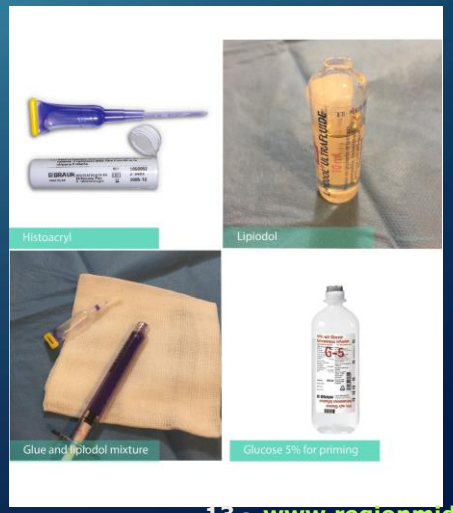
Embolization material

- Coils - Macrocoils and
- microcoils
- Microplug (MVP)
- Onyx, Glue

VV performed under GA/LA
Direct puncture with thin needles

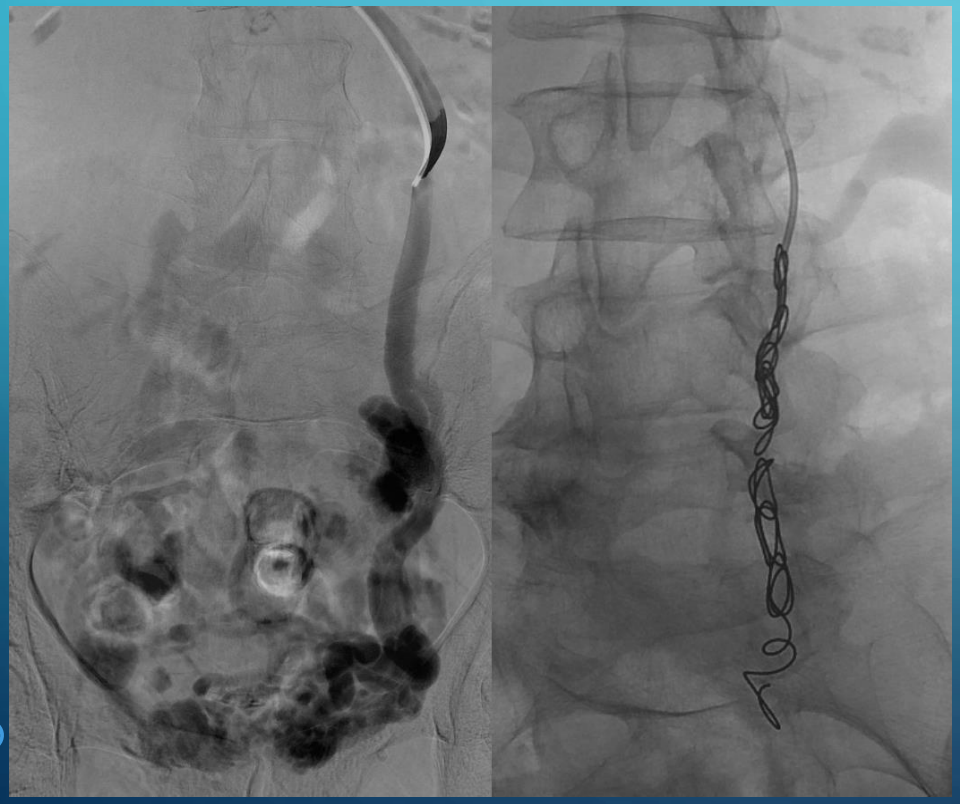


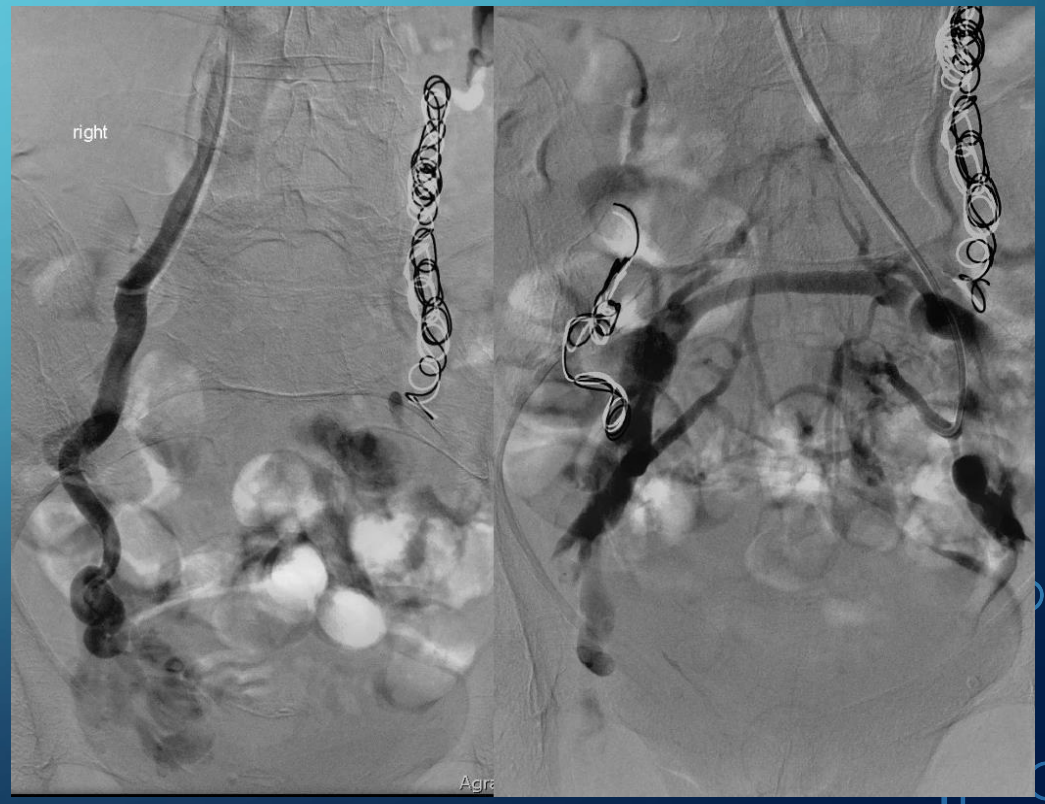
Foam sclerotherapy - STS, Polydocanol, Alcohol etc



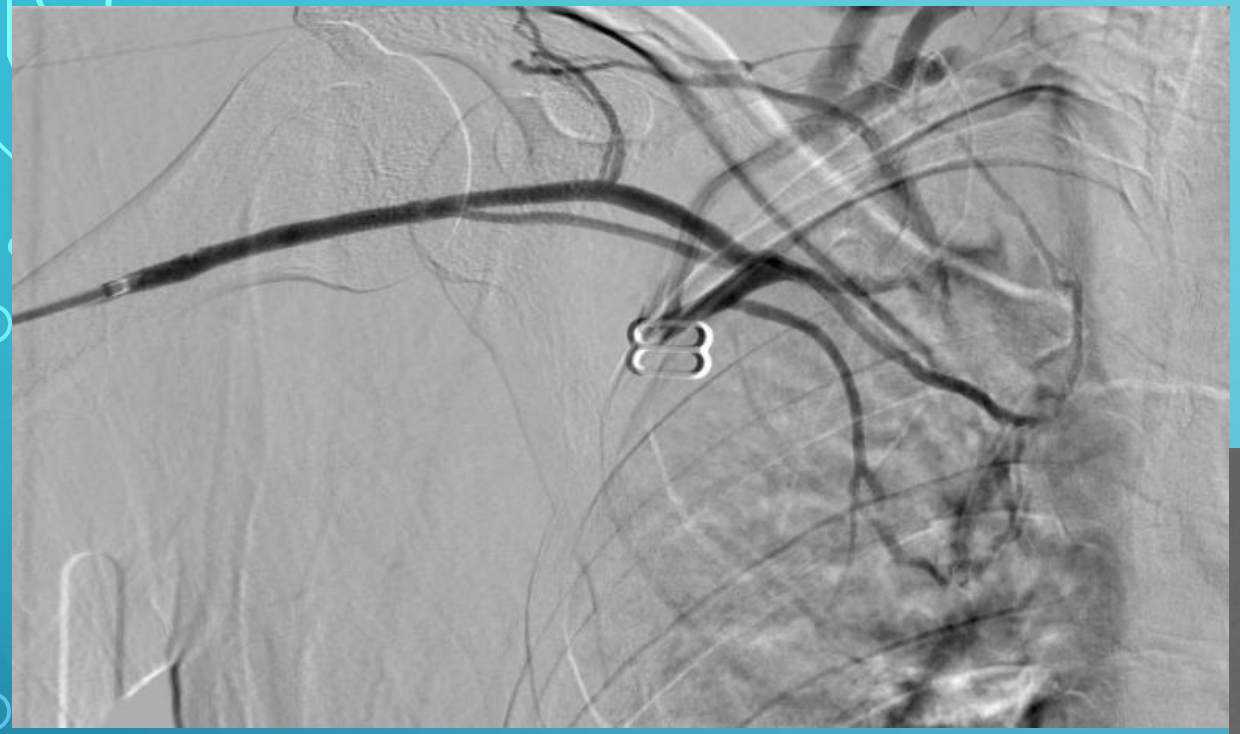
PCS - EMBOLIZATION

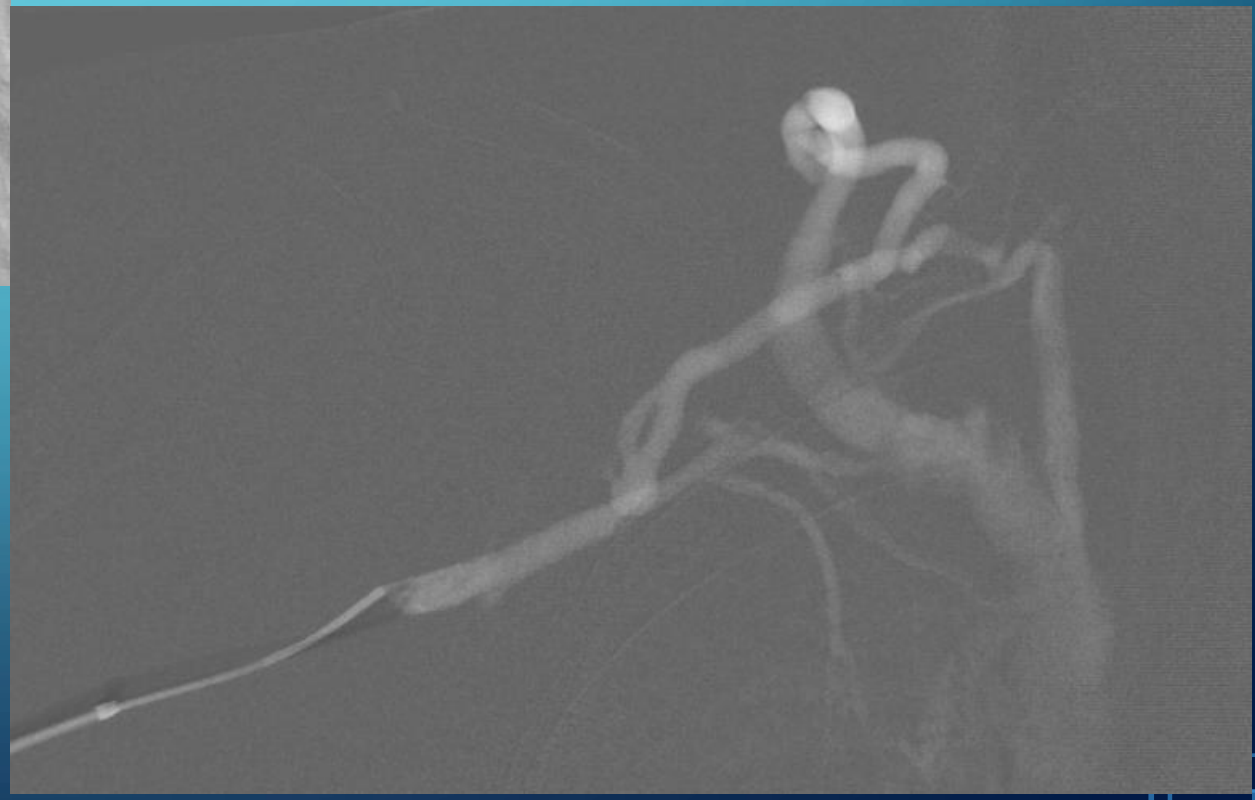
- Bilateral or in some cases can be unilateral
- Femoral, brachial/basilic or jugular approach
- Phlebography confirms reflux, dilatation, tortuousity etc.
- Embolization of bilateral ovarian and sometimes internal iliac veins.



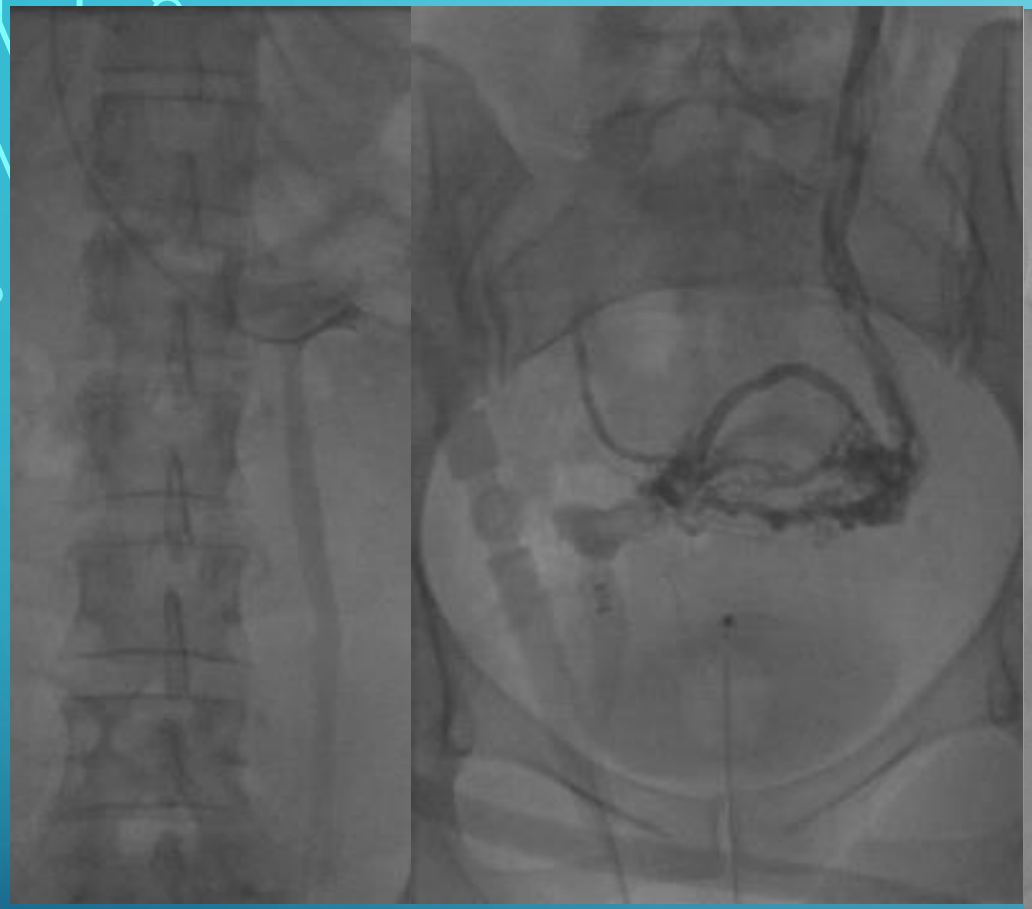


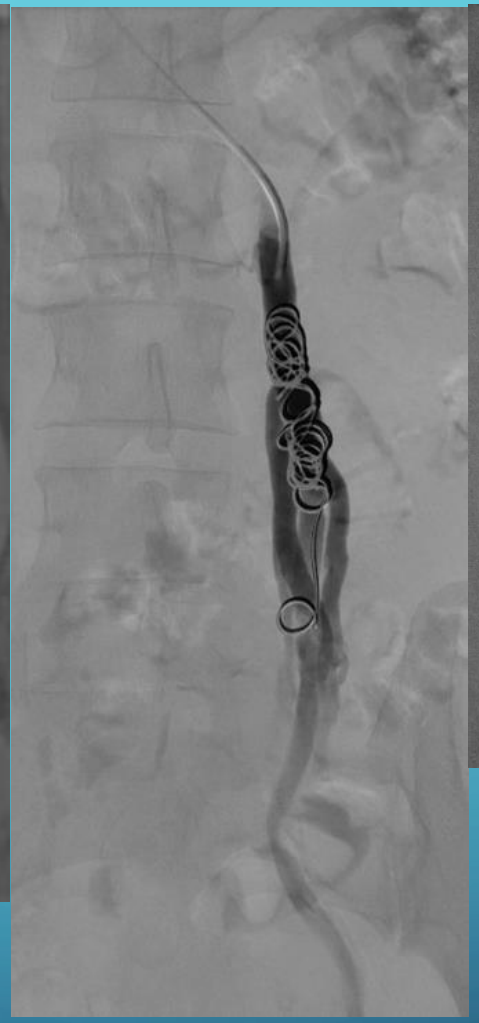
PCS - EMBOLIZATION

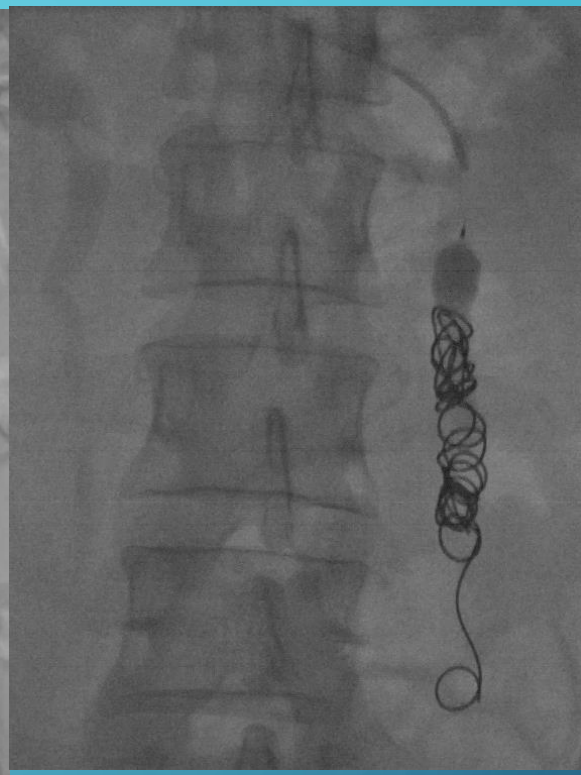


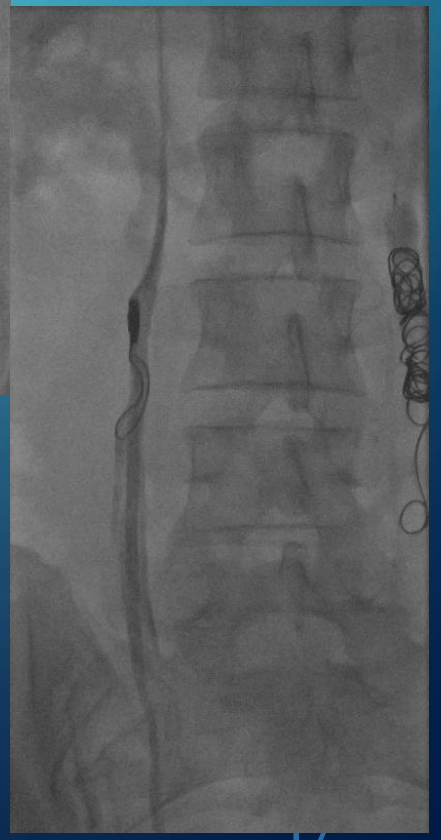


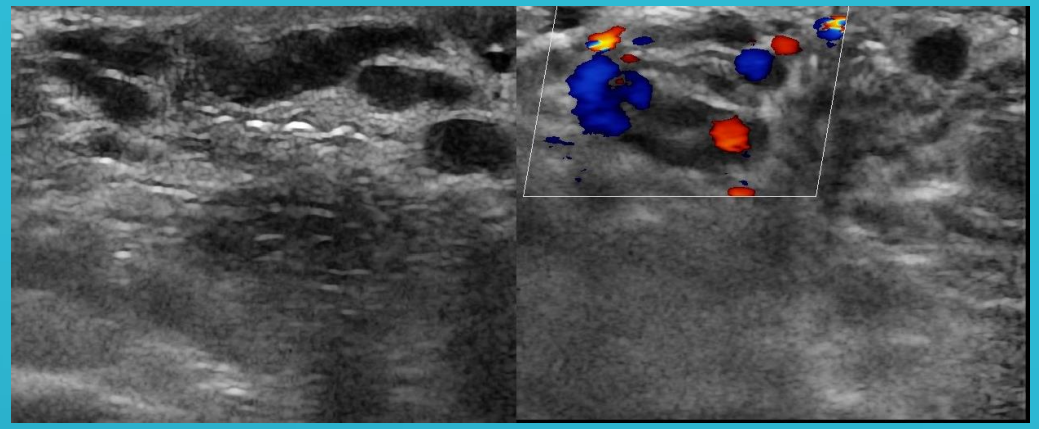
PCS - EMBOLIZATION



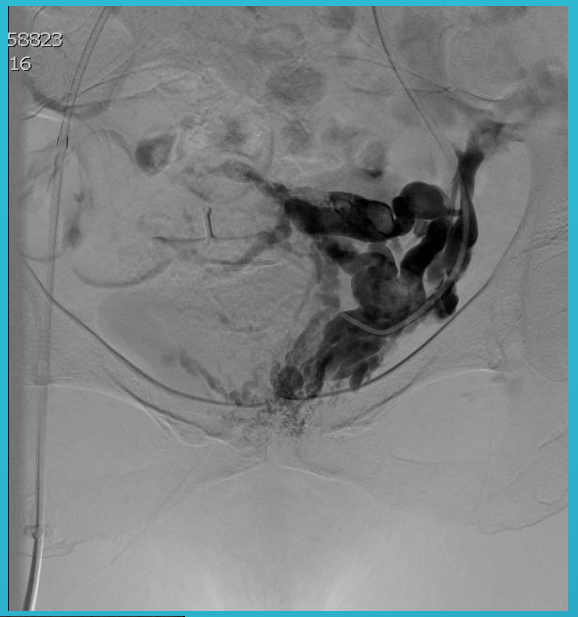


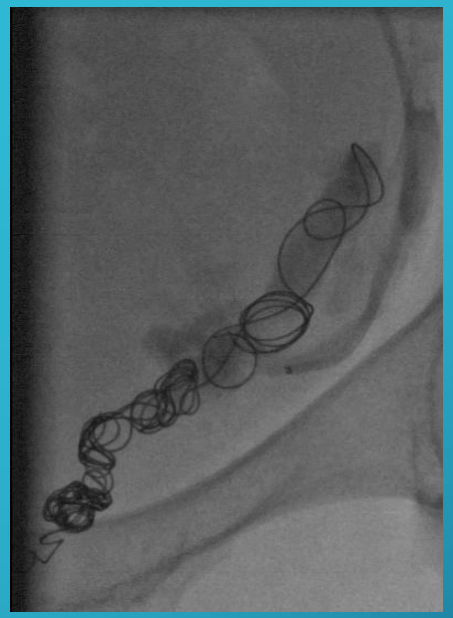






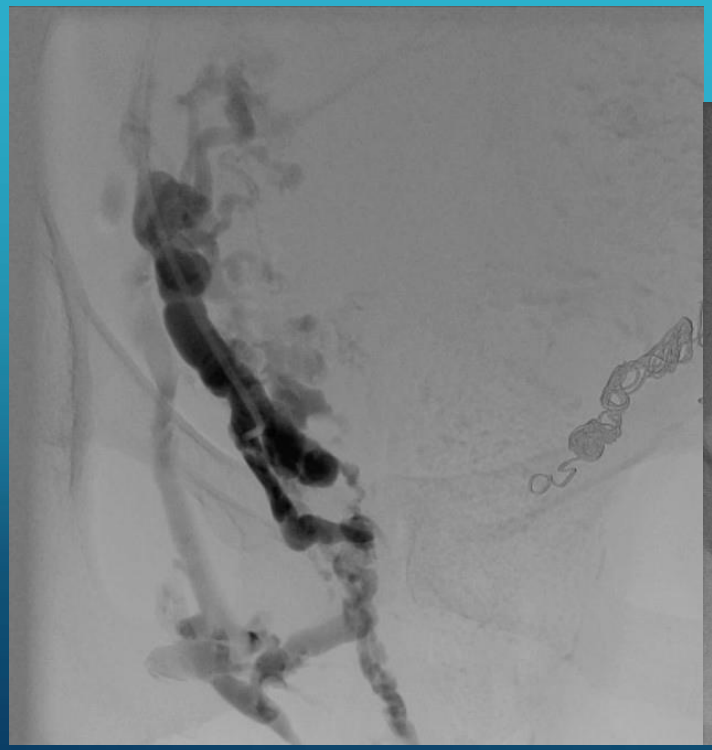
Presence of tortuous and dilated vulval veins bilaterally (sag. & axial)

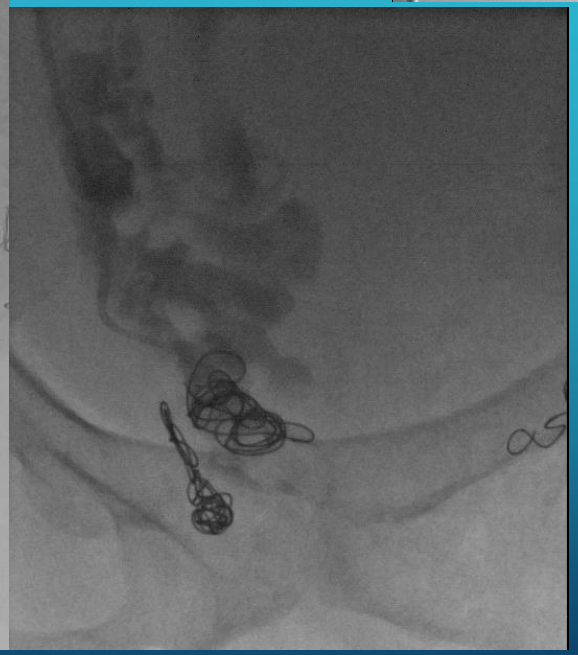




Presence of tortuous and dilated pelvic venous plexuses on left side communicating with int. iliac veins (left)

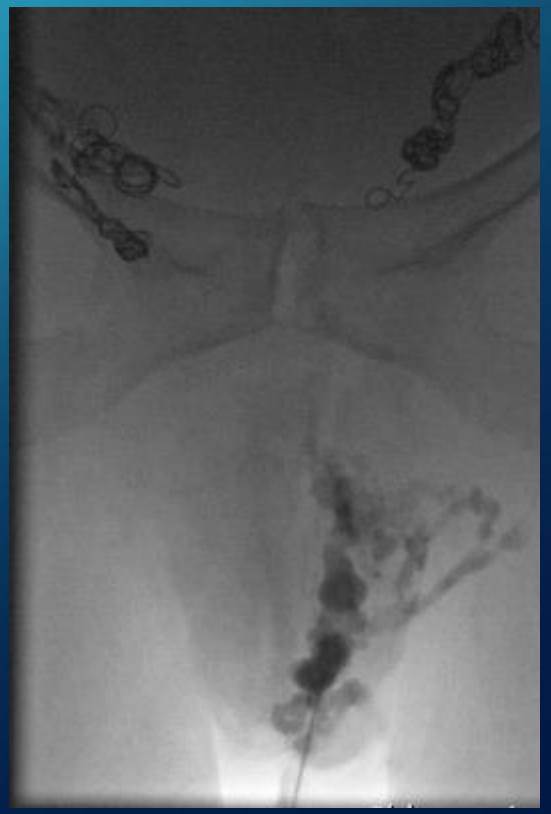
Embolization with Azur macrocoils and Onyx

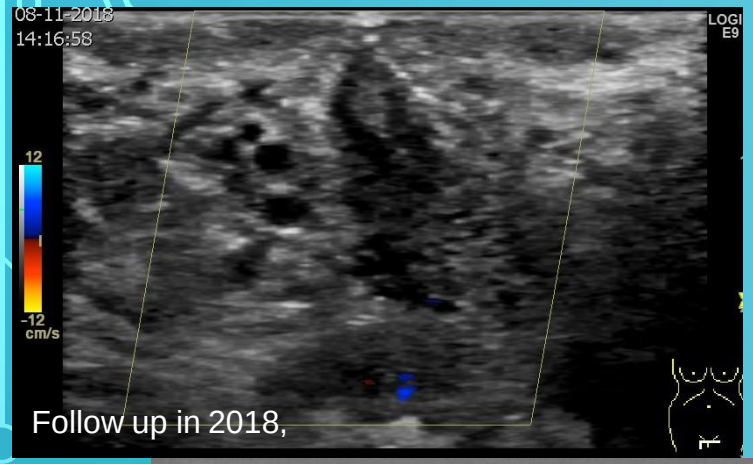




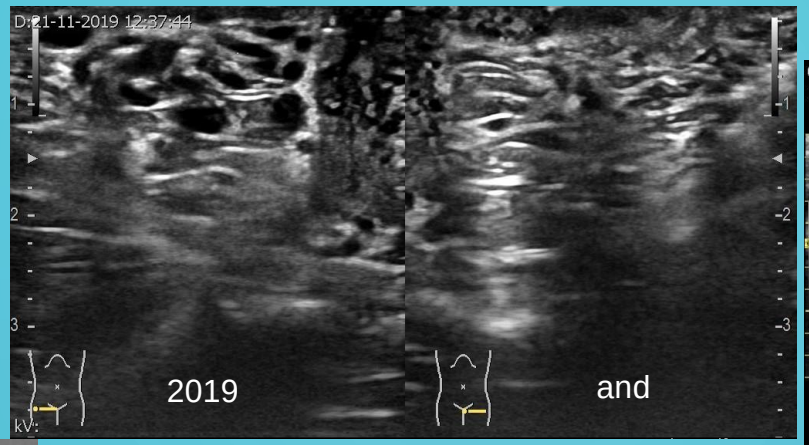
Presence of tortuous and dilated pelvic venous plexuses on right side communicating with int. iliac veins (right)
Embolization with Azur macrocoils and Onyx

Simultaneous Sclerotherapy of the left sided VV with STS (Sotradecol) foam.



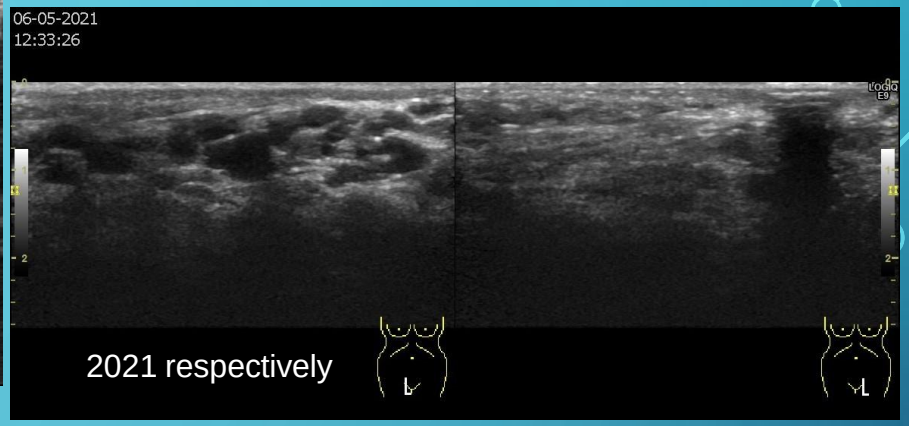


Follow up in 2018,

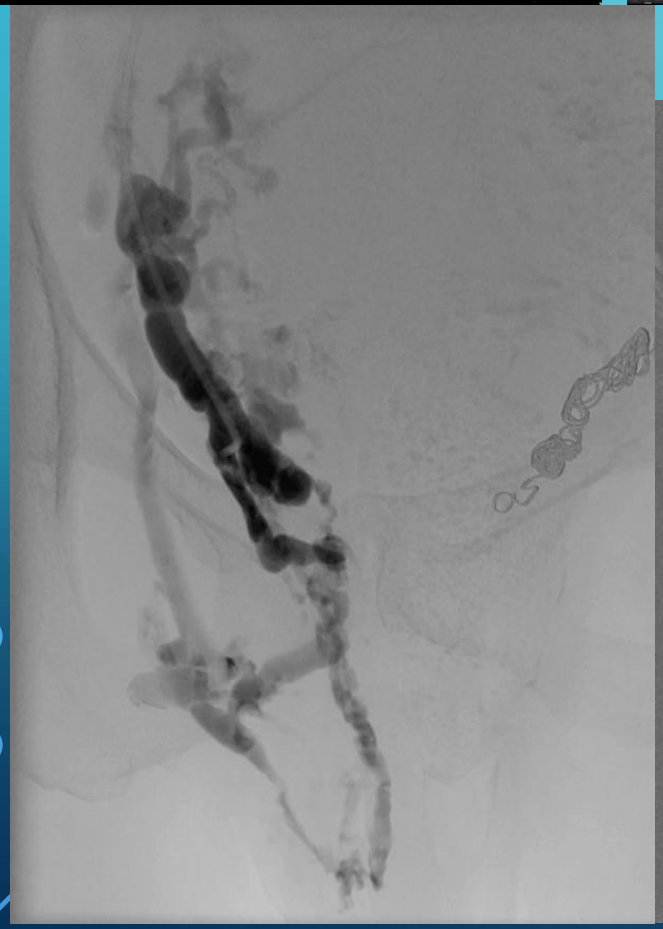


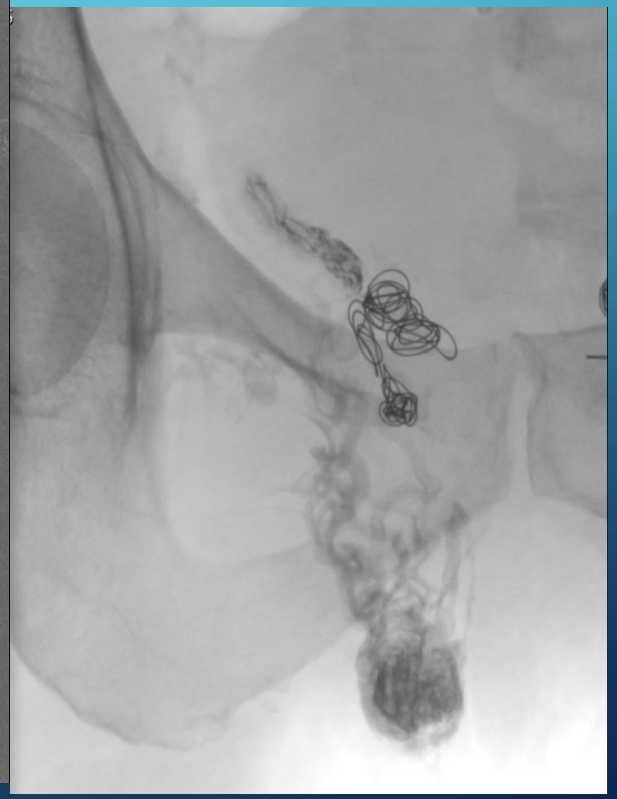
2019

and

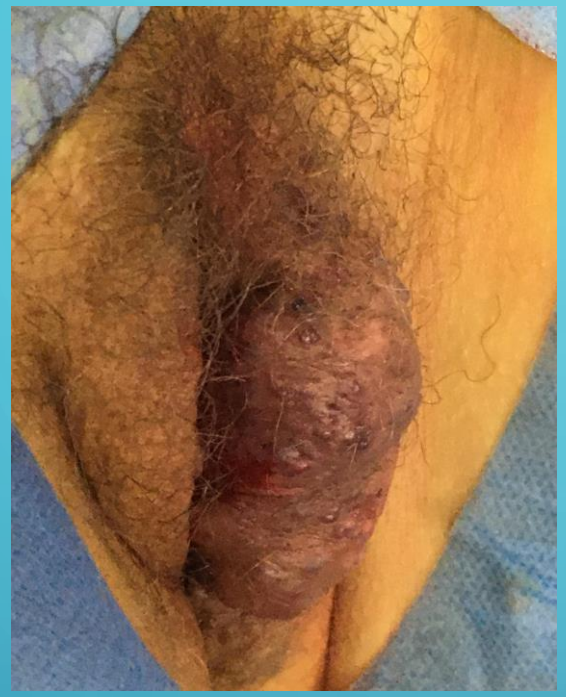


2021 respectively





Foam sclerotherapy of the right sided VV with STS (Sotradecol).



Foam sclerotherapy of the left sided VV with STS (Sotradecol).

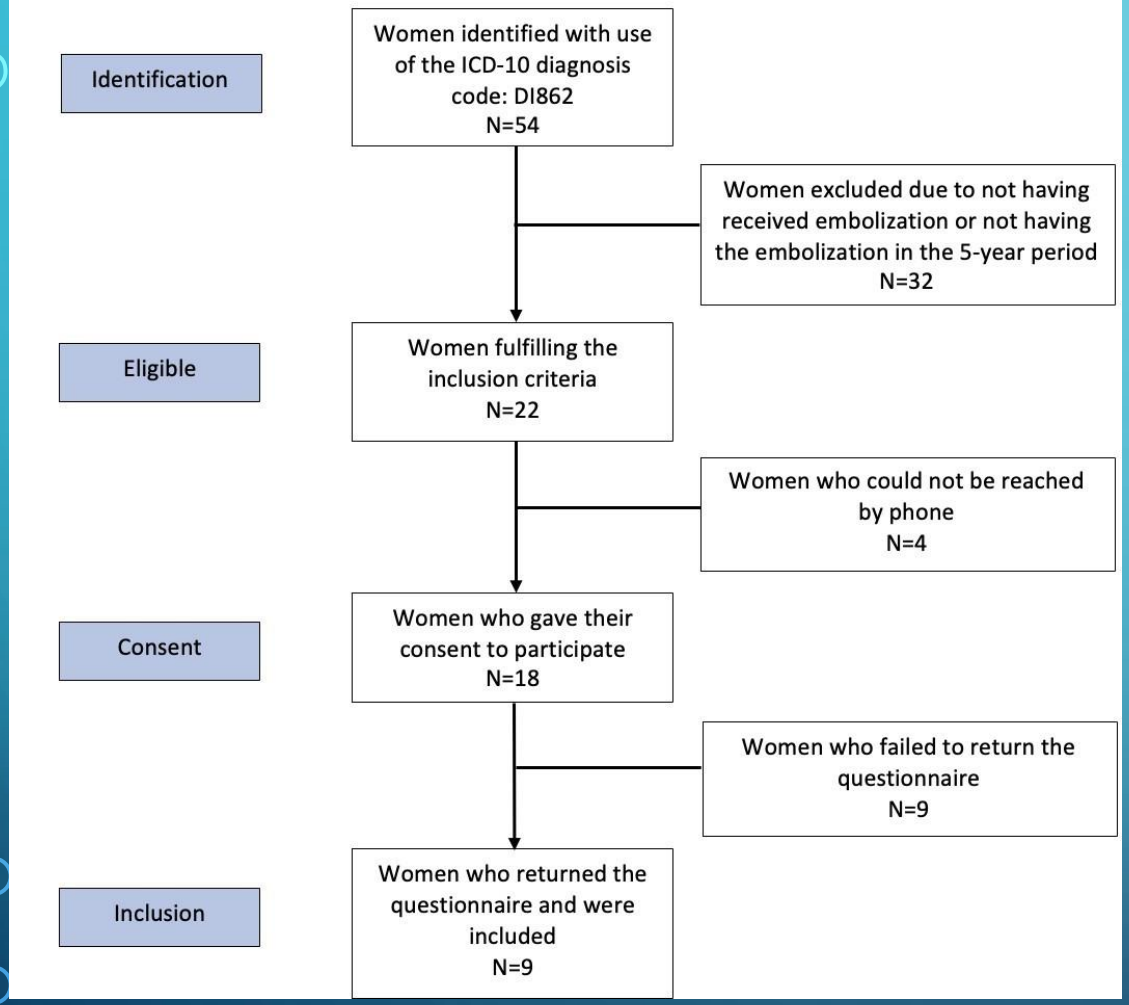
Follow up at 6 weeks after last treatment

RESULTS FROM TREATING PELVIC CONGESTION SYNDROME AND VULVAR VARICOSITIES - THE PAST 5 YEARS AT AARHUS UNIVERSITY HOSPITAL

- By Emma Cordsen Buchhave, MD and Frederik Ehlern Larsen, MD
- Supervisors:
- Mette Meinert, MD, PhD, Department of Gynecology and Obstetrics, AUH *Arindam Bharadwaz*, MD, EBIR, Department of Radiology, AUH

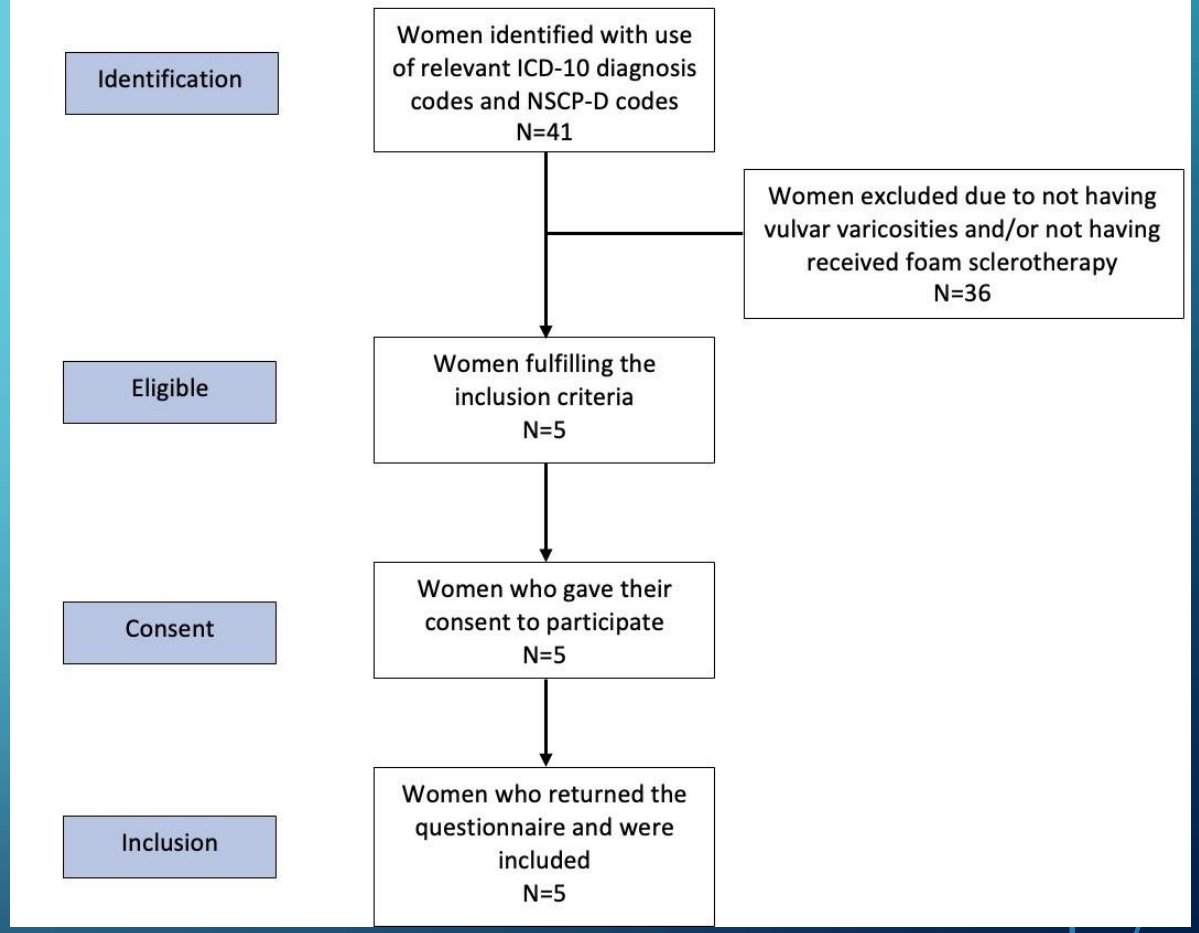
STUDY DESIGN AND RECRUITMENT

Figure 1: Flowchart - recruitment of women with PCS treated with embolization



01.08.2014 - 01.08.2019

Figure 2: Flowchart - recruitment of women with vulvar varicosities treated with foam sclerotherapy



01.01.2016 - 15.01.2021

RESULTS – EMBOLIZATION

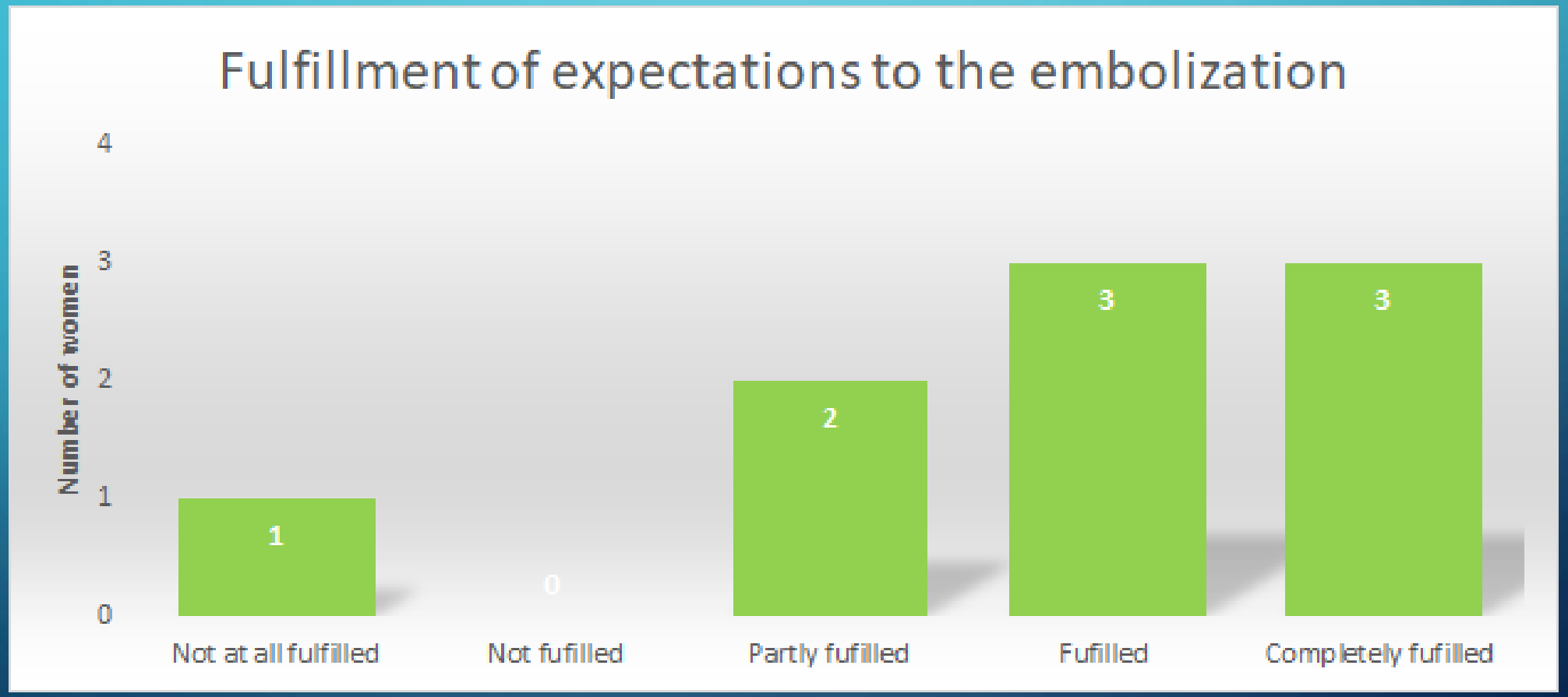
Table 3
Outcomes for the group of women with PCS treated with embolization (N=9)

	Before embolization	After embolization	Difference	*P*-value
Pain (NRS[a])	7.11 ± 1.54	3.11 ± 3.14	–4.00 ± 3.74	0.01
Dyspareunia (NRS[a])	5.33 ± 3.50	1.56 ± 1.67	–3.78 ± 3.07	0.01
Illness' impact on life quality[b]	8.11 ± 1.45	2.89 ± 3.44	–5.22 ± 2.95	0.00

Values are listed as a mean ± SD. Differences are calculated as: value after treatment - value before treatment.

Mean time since treatment: 2.31 years (SD ± 1.36)

RESULTS - EMBOLIZATION



RESULTS - FOAM SCLEROTHERAPY

Table 4
Outcomes for the group of women with vulvar varicosities treated with foam sclerotherapy (N=5)

	Before foam	After foam	Difference	*P*-value
Pain (NRS[a])	6.40 ± 2.88	0.80 ± 1.30	–5.60 ± 2.07	0.00
Dyspareunia (NRS[a])	4.00 ± 4.62	2.75 ± 4.86	–1.25 ± 3.95	0.57
Quality-of-life[b]	4.40 ± 3.05	9.40 ± 0.89	5.00 ± 2.55	0.01
Limitations in physical activity[c]	6.60 ± 2.70	0.60 ± 1.34	–6.00 ± 2.00	0.00
Sensation of heaviness in vulvar area[d]	3.80 ± 1.64	1.60 ± 0.89	–2.20 ± 1.30	0.02
Sensation of bulging in vulvar area[d]	4.80 ± 0.45	1.60 ± 0.89	–3.20 ± 0.84	0.00

Values are listed as a mean ± SD. Differences are calculated as: value after treatment - value before treatment.

Mean time since treatment: 1.5 years (SD ± 1.1)

Satisfaction of treatment outcome measured on a scale from 0 to 10: 9.80 (SD ± 0.45)

CONCLUSION AND LIMITATIONS

Conclusion:

- Pain ⇓
- Quality of life ⇑
- Dyspareunia after embolization ⇓

Limitations:

- Small study
- Risk of recall bias
- Effect of foam sclerotherapy or embolization?

* The largest series so far of reports treating 7 cases of VV

Ninia JG. Treatment of vulvar varicosities by injection-compression sclerotherapy. Dermatol Surg. 1997 Jul;23(7):573-4; discussion 574-5. doi: 10.1111/j.1524-4725.1997.tb00689.x. PMID: 9236876.

Castenmiller, P. H., de Leur, K., de Jong, T. E. & van der Laan, L. *Clinical results after coil embolization of the ovarian vein in patients with primary and recurrent lower-limb varices with respect to vulval varices*. *Phlebology* **28**, 234-238, doi:10.1258/phleb.2012.011117 (2013).

KEY POINTS – PCS

1. PCS is the result of dilated pelvic veins resulting in chronic pelvic pain (CPP).
2. Other causes of CPP should be ruled out.
3. Diagnosis of PCS should be based on patient history as well as clinical and imaging findings.
4. Clinical evidence regarding the efficacy of medication therapy for management of PCS remains limited and available therapies have not been shown to produce long-term improvement.
5. Endovascular treatment seems superior to surgery.

Bendek B, Afuape N, Banks E, Desai NA. Comprehensive review of pelvic congestion syndrome: causes, symptoms, treatment options. Curr Opin Obstet Gynecol. 2020 Aug;32(4):237-242. doi: 10.1097/GCO.0000000000000637. PMID: 32487799.

KEY POINTS – VV

1. VV is primarily a clinical diagnosis and are visible on inspection.
2. Diagnosis is supplemented by Imaging.
3. VV can be isolated or associated with PCS or leg varices.
4. VV can cause significant symptoms, both physical and psychological.
5. Sclerotherapy seems to be superior to surgery.

Bendek B, Afuape N, Banks E, Desai NA. Comprehensive review of pelvic congestion syndrome: causes, symptoms, treatment options. Curr Opin Obstet Gynecol. 2020 Aug;32(4):237-242. doi: 10.1097/GCO.0000000000000637. PMID: 32487799.

THANK YOU