The
Ehlers
Danlos
Society™

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OB/GYN and EDS/HDS

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OB/GYN and EDS/HDS

- Review gynecologic and obstetric issues seen with EDS/HDS
  - Gynecologic concerns including pain and organ prolapse
  - Puberty/adolescence
  - Sexually active woman
  - Fertility
  - Menopause

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OB/GYN and EDS/HDS

- Obstetrical Management
  - Miscarriage
  - During Pregnancy
    - Symptoms
    - Complications
  - Delivery
  - Postpartum

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Puberty

- Symptoms of EDS can become worse with puberty, or can begin at puberty
  - Hugon-Rodin 2016 series of 386 women with hypermobile type EDS.
    - 52% who had prepubertal EDS symptoms (chronic pain, fatigue) became worse with puberty.
    - 17% developed symptoms of EDS with puberty

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GYN Issues EDS/HDS

- **Bleeding disorders –** the most common gyn symptom
- **Menorrhagia** – heavy bleeding 33-75%, worst in vEDS
  - Weakness in capillaries and perivascular tissue
  - Abnormal interaction between platelets and collagen
- **Dysmenorrhea** – painful menses 73-93%
  - Usually caused by prostaglandins
    - Made in lining of uterus, cause muscles and blood vessels of uterus to contract

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GYN Bleeding Treatment

- **Dysmenorrhea**
- **NSAID** – non-steroidal anti-inflammatory drug, target prostaglandins
- Works best if start at very onset of menses
- Can help with nausea, diarrhea
- Alternative treatment options include **Vitamin B1, magnesium supplements, acupuncture**

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Hormonal therapy

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GYN Bleeding Hormonal Treatment

- **Oral Contraceptive Pill or Progesterone only medication** (mini pill, antigonadotropic agents), **IUD with progesterone**
- Bleeding decreases, can be suppressed
- Dysmenorrhea improves
- Hugon-Rodin
  - EDS symptoms improved in 15% on OCP, 25% on progesterone only medication
  - EDS symptoms worse in 25% on OCP who already had cyclic worsening of EDS each perimenstrual period, improved in 15%

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Vulvodynia/Dyspareunia

- Vulvodynia: chronic pain /discomfort of the vulva for which no obvious etiology can be found.
- Incidence 32-77%. Always have a history of dysmenorrhea
- Etiology- peripheral sensitization of the vulva or generalized urogenital, pelvic and/or central nerve nervous sensitivity
- Severe vulvar edema during/after intercourse
- Persistent genital arousal syndrome

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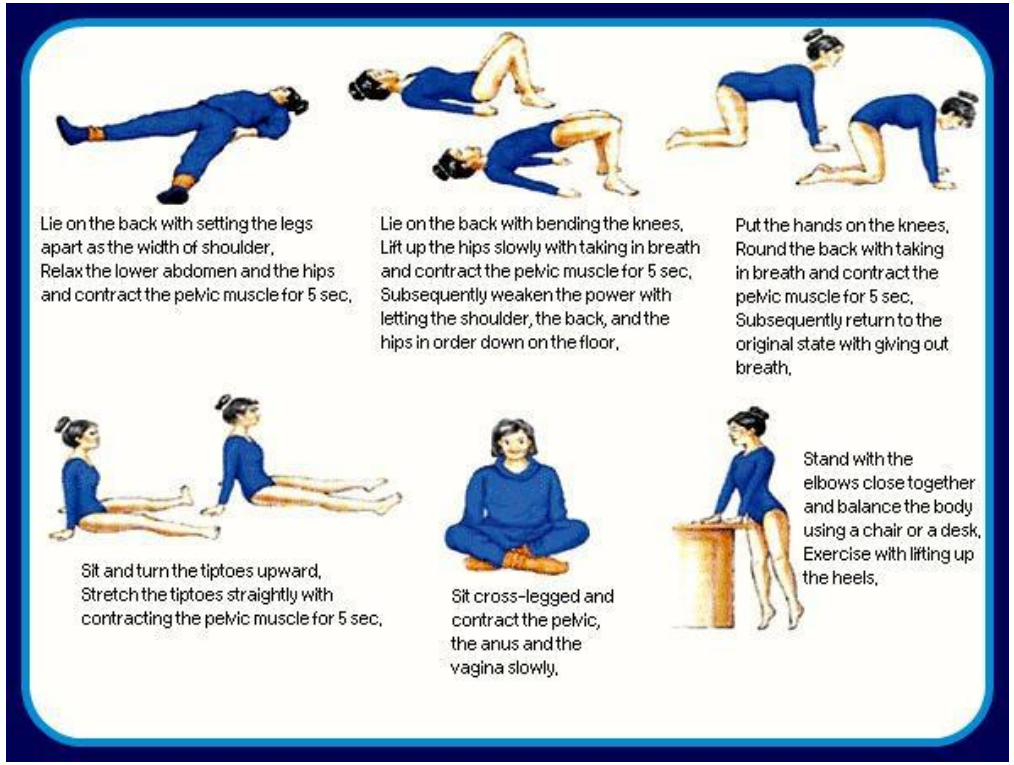
Vulvodynia/Dyspareunia: Treatment

- Skin care
- Physical therapy
- Psychotherapy
- Cognitive behavior therapy
- Exercises: Kegel/reverse Kegel
- Medications



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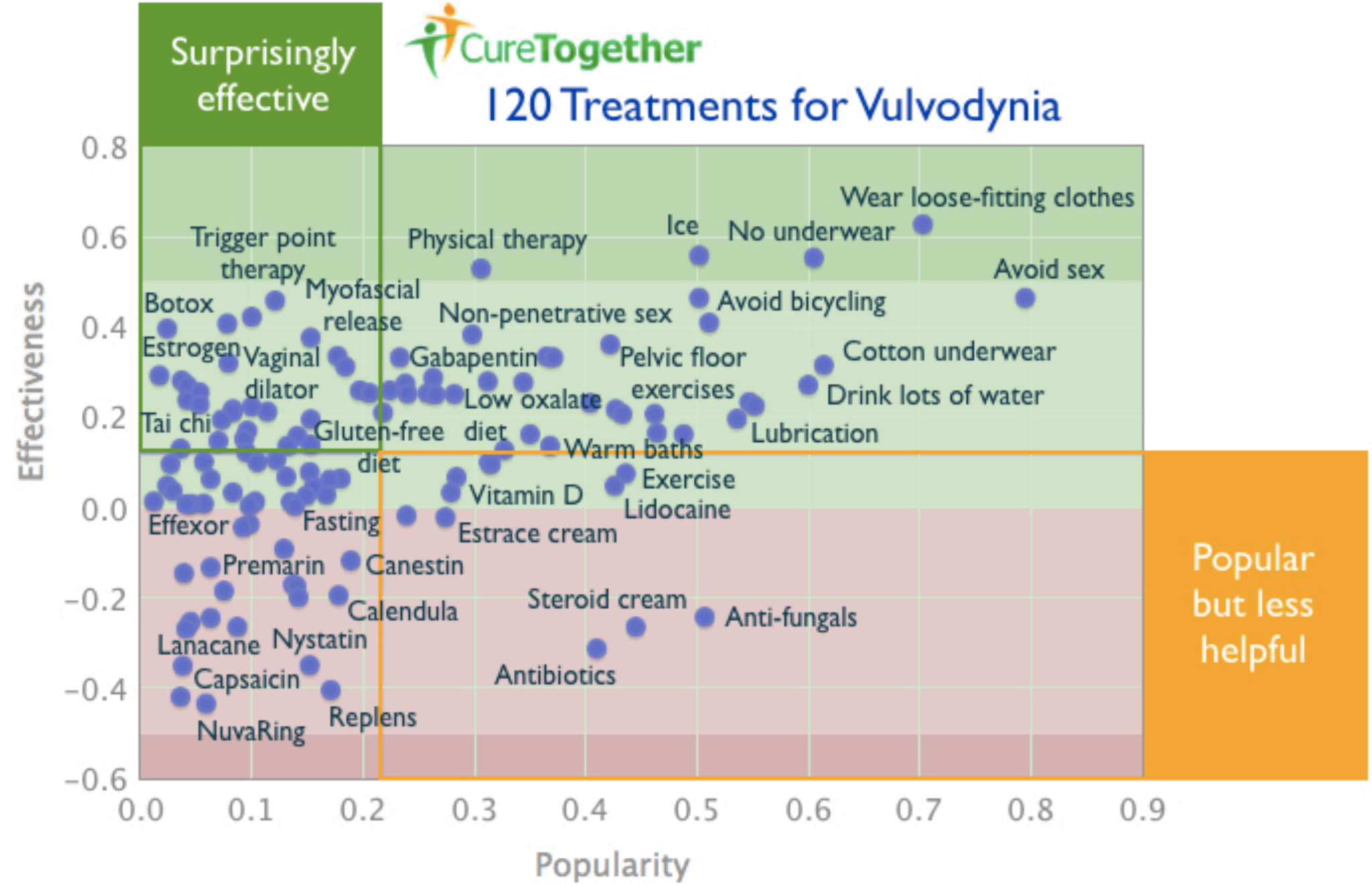

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Other Gyn issues

- **Endometriosis**
- 6%-23%
- **Uterine myomas**
- 5-9%
- **Infertility**
- hEDS: No difference from general population (majority of studies) to 48%
- **Menopause**
- Improvement of EDS symptoms in 22%
- HRT used in 42%. 15% of these women founded symptoms improved.

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Obstetrics

- Physiologic/hormonal changes
- Increased levels of relaxin hormone, can exacerbate pre-existing joint laxity and pain in hEDS.
  - Three times more likely to require treatment for **pelvic girdle pain and instability** than the general population.
  - **Spine and joint pain**, low back pain, pelvic pain in sacroiliac joints, midline groin pain radiating to lower abdomen and hips (pubic symphysis dysfunction).
  - **Gastrointestinal reflux** common in pregnancy, more common in hEDS. Antacids, avoid spicy foods, opiods.
  - **Varicose veins** in legs, vulva during pregnancy. Treat with compression hose.

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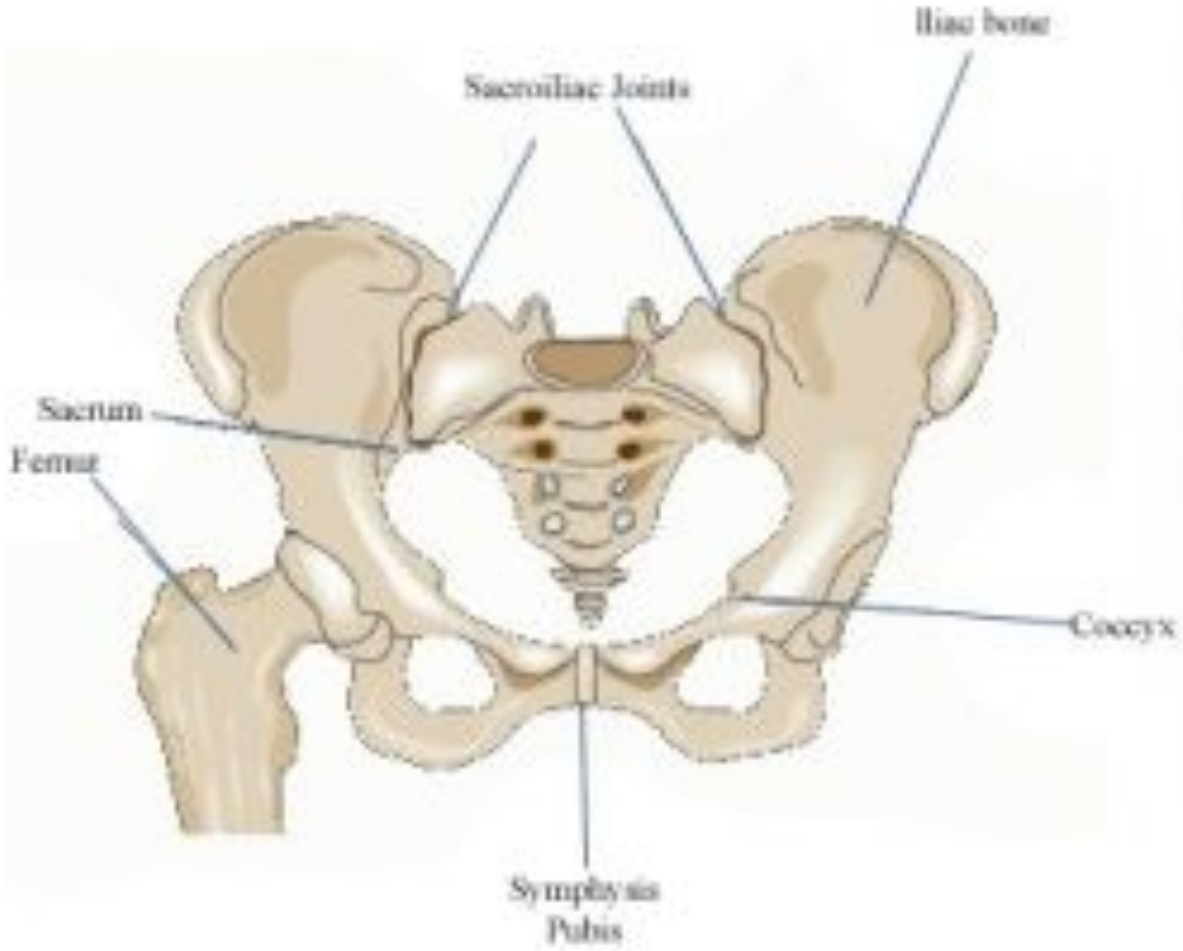
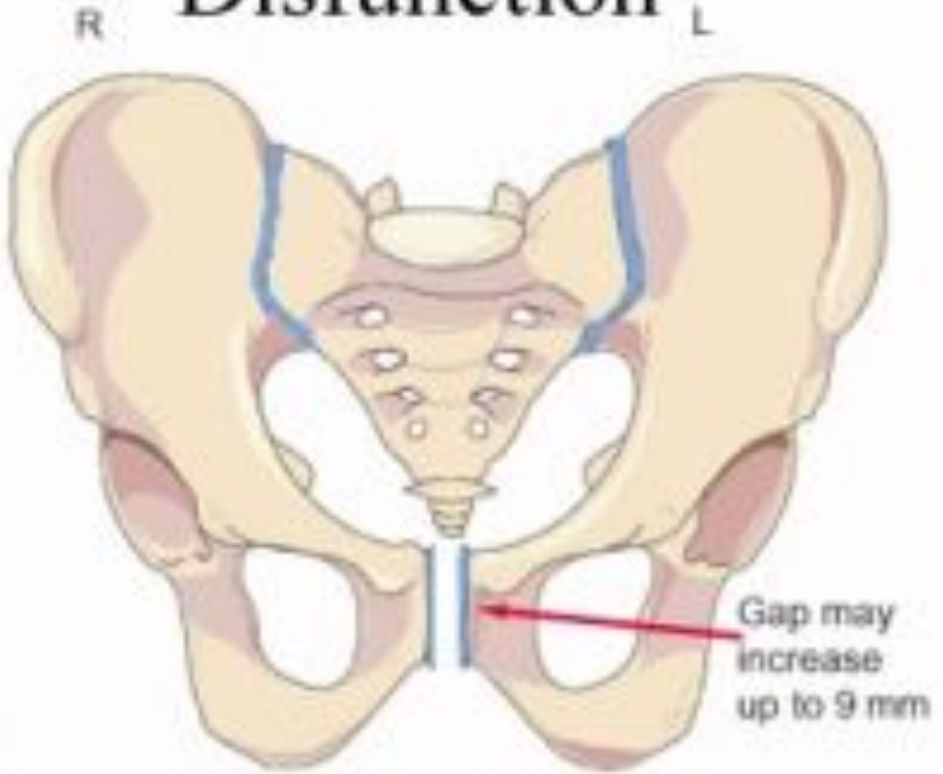
Normal Pelvis



Symphysis Pubis Disfunction



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Obstetrics: POTS

- Postural orthostatic tachycardia syndrome
  - Heart rate increases 30 bpm or to greater than 120 bpm when moving from supine to upright position.
- Pregnancy cardiovascular changes such as peripheral venous blood pooling, leading to reduced diastolic blood pressure, may exacerbate POTS symptoms:
  - Dizziness, nausea, palpitations, fatigue, fainting.
  - Can worsen with pain of labor

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Obstetrics: POTS

- Kanjwal studied 22 POTS pts in pregnancy, 2 hypermobile. 53% improved, 31%worse,4/7 only during first trimester. Third trimester greater fluid retention is of benefit.
- Treatment:
  - Adequate salt and fluid intake
  - Adequate anesthesia for labor
  - Avoid Valsalva during second stage of labor

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Obstetrics

- Rate of miscarriage (loss of pregnancy in first 12 weeks) similar to population risk in most studies. Several studies have noted higher rates, hEDS 28%
- Loss of twin pregnancies. Lind reported 4 twin pregnancies, 3 late SAB, one 29 week delivery.

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Obstetrics: PROM

- Increased risk for PROM- Premature rupture of membranes, which can lead to preterm delivery, before 37 weeks gestational age.
  - Amnion: 5 layers, including a layer mainly made up of type 1 & 2 collagen fibrils.
    - Reduced number of fibrils in PROM.
    - Theory in EDS, altered fibrils lead to PROM.

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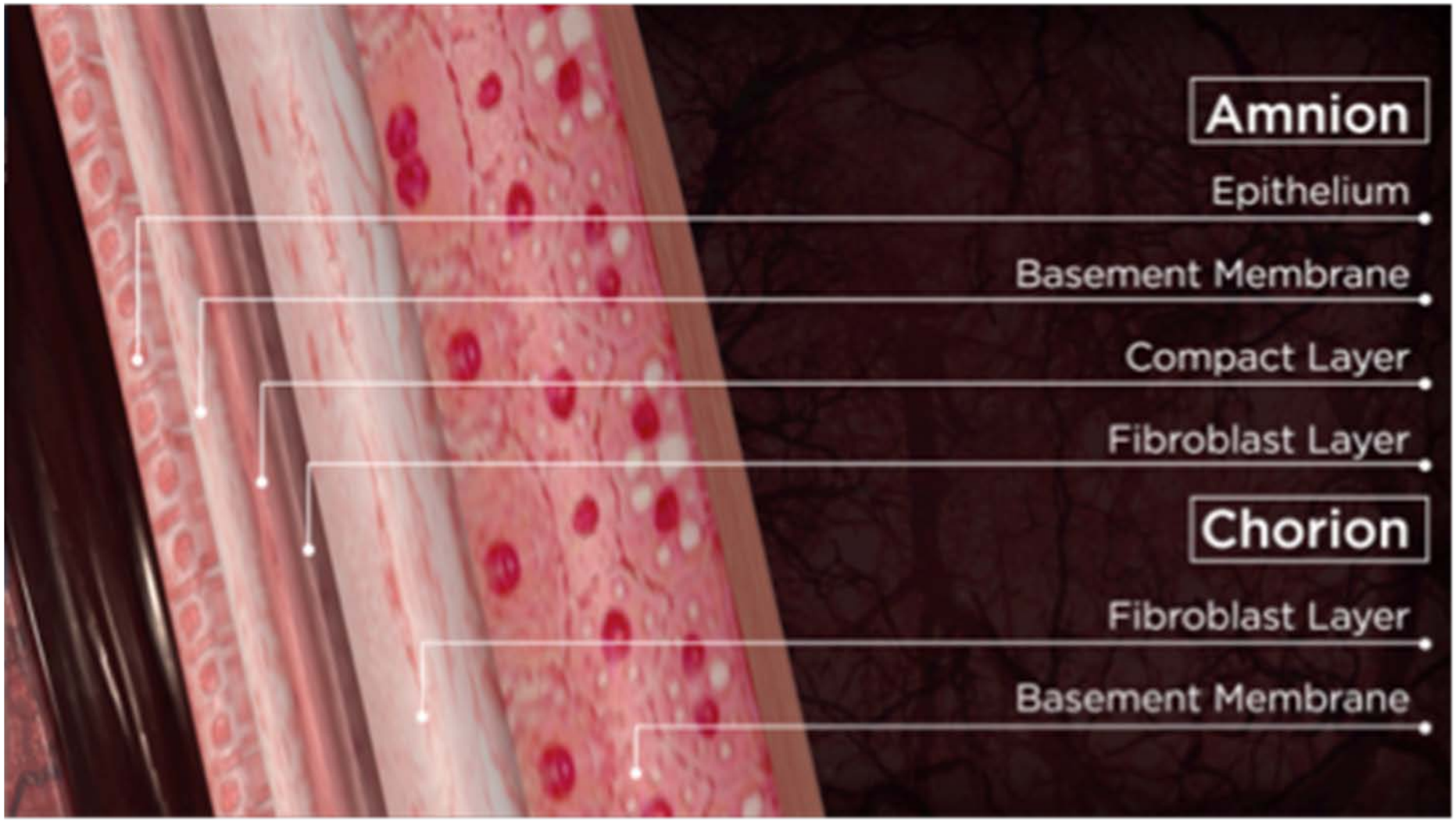

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Obstetrics: PROM

- Lind EDS 66pts, 264 pregnancies and 33 nonEDS pts, 107 pregnancies affected with EDS (new mutation46%, paternal 54%).
  - EDS mother, PROM 20% (baby also EDS, 35%)
  - nonEDS mother (baby EDS), PROM 50%
  - Did not see incompetent cervix in this series
  - Has been reported, particularly in cEDS

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Obstetrics PROM

- Castori: 82 hEDS women, 93 pregnancies
- 11% PTD, likely PROM
- Hurst: Large survey, 25% preterm birth rate
- Hugon-Rodin: 6% PTD

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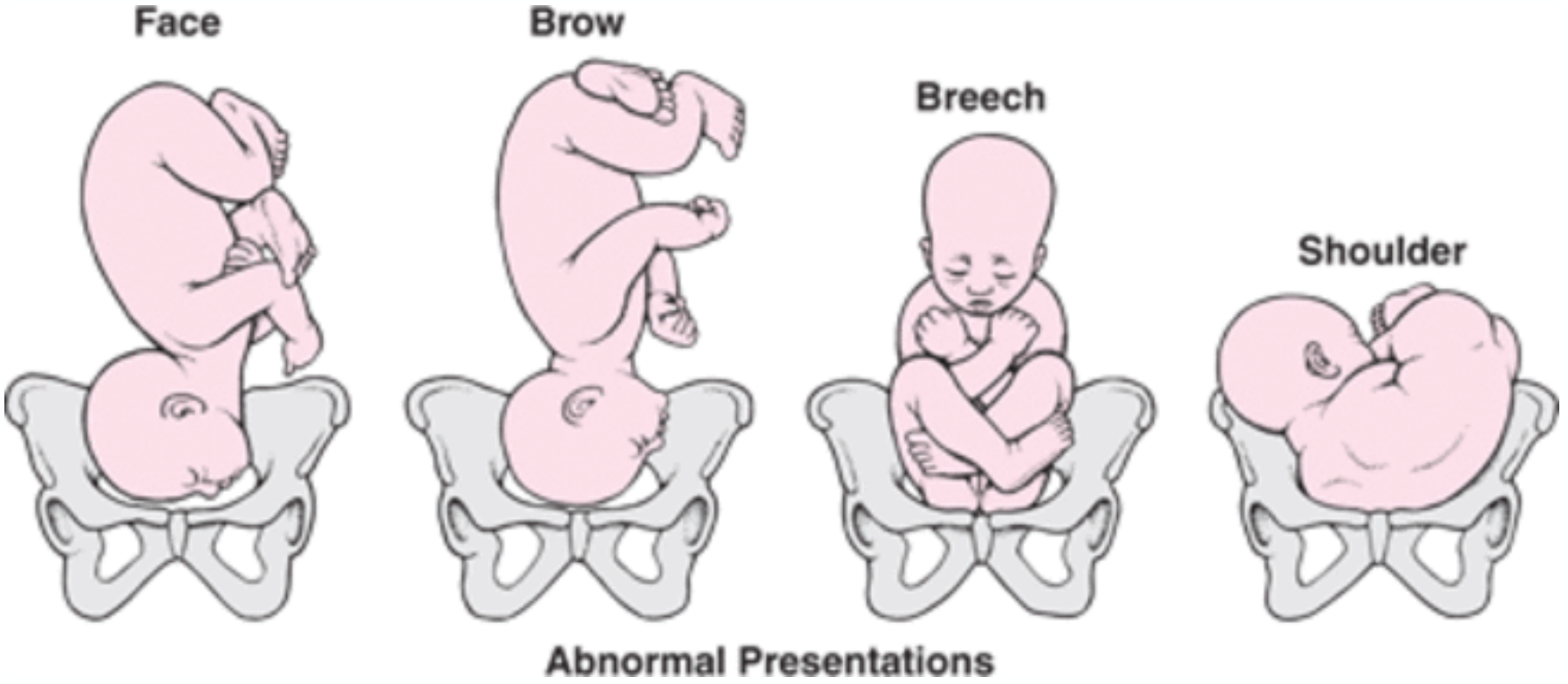
Obstetrics: Fetal presentation

- Breech presentation 8% vs 3% in general population
- Lind study: Face presentation, Brow presentation in 5/46, all affected infants.



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Obstetrics

- Mode of delivery
- Vaginal
- Avoids abdominal surgery
- Risk of perineal injury during second or third stage of labor, or episiotomy extension
- Risk of wound opening or slow healing, scar atrophic or keloid
- Precipitous delivery, < 4 hours, 36%

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Obstetrics: Labor

- Knoepp found women with hypermobility experienced:
  - Less operative intervention in the second stage of labor.
  - Less likely to have prolonged second stage (not statistically significant).
  - Less likely to have anal sphincter laceration.
- Joint laxity may allow for more easy passage of fetal head through the pelvis.
- Hypermobility did not increase the relative odds for any pelvic floor disorders 5-10 years after delivery.

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Obstetrics

- Risk of hemorrhage 19%
- During labor 10%
- Postpartum 5%
- Use of DDAVP for uterine atony

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Obstetrics: Cesarean Delivery

- Avoids labor if elective
- Avoid pelvic floor injury
- Wound may heal slowly or break down. Abnormal scar formation 46%

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Obstetrics: Cesarean Delivery

- Recommended for
- Fetal malpresentation
- Excessive joint pain
- Preserve pelvic floor? No data to support this.

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Obstetrics: Pelvic Floor

- Risk of Pelvic Prolapse 15%-30%
- Weakened pelvic floor can lead to cystocele, bladder distention, stress urinary incontinence (33% in one study)
- Pelvic floor exercises may help
- Prolapse can occur in nulliparous patients, even in teen years.

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Obstetrics: Management of Delivery

- Management of vaginal delivery/C Section
- “Gentle” pushing, that is spontaneous pushing rather than directed pushing
- Appropriate positioning, avoiding overextending hips. Risk of injury may be greater with regional anesthesia, eliminating pain as warning sign.
- Gentle handling of tissue.
- Retention sutures, left in place for 2 weeks. Non tension, deep placement.
- Permanent sutures rather than dissolving sutures.

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Obstetrics: Classical EDS

- PROM
- Cervical incompetence
- Breech presentation
- Episiotomy extensions, perineal tears
- Pelvic prolapse leading to fecal andor urinary incontinence, treatment problematic

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Obstetrics: Vascular EDS

- Risk of arterial dissection, rupture during pregnancy, in labor, post partum.
- Pregnancy specific complications in 50%:
  - Uterine rupture
  - Hemorrhage per, during and post delivery
  - PROM, PTD
  - Severe peritoneal tears

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Obstetrics: Vascular EDS

- Murray 35 women, 76 pregnancies
- 62% vaginal delivery, 33% routine Cesarean section, 5 emergency Cesarean section,
- 48% uncomplicated deliveries (vaginal and CS).
- 22% PTD, 21% 3rd/4th degree laceration(10x population rate), 11% hemorrhage, 4% previa, 3% abruption, one intraoperative bladder and venous tear.

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Obstetrics: Vascular EDS

- Murray, cont.
- Vascular events:
- 4 Lethal: 1 iliac rupture after fall at 33 weeks gestation, 2 aortic rupture in labor, 1 aortic rupture 7 days post Cesarean delivery
- 3 Nonlethal: 1 coronary artery dissection at term. 1 splenic artery dissection 6 days post SVD, 1 multiple artery dissection 7 days post SVD
- 2 Uterine ruptures, 1 antepartum, one intrapartum

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Obstetrics: Vascular EDS

- Murray cont.
- Conclusion: For women who do not have vEDS complications before pregnancy, pregnancy itself does not appear to add further risk of death.
- Calculated mean age of death is 53 years old, whether had pregnancies or not.

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Obstetrics: Vascular EDS

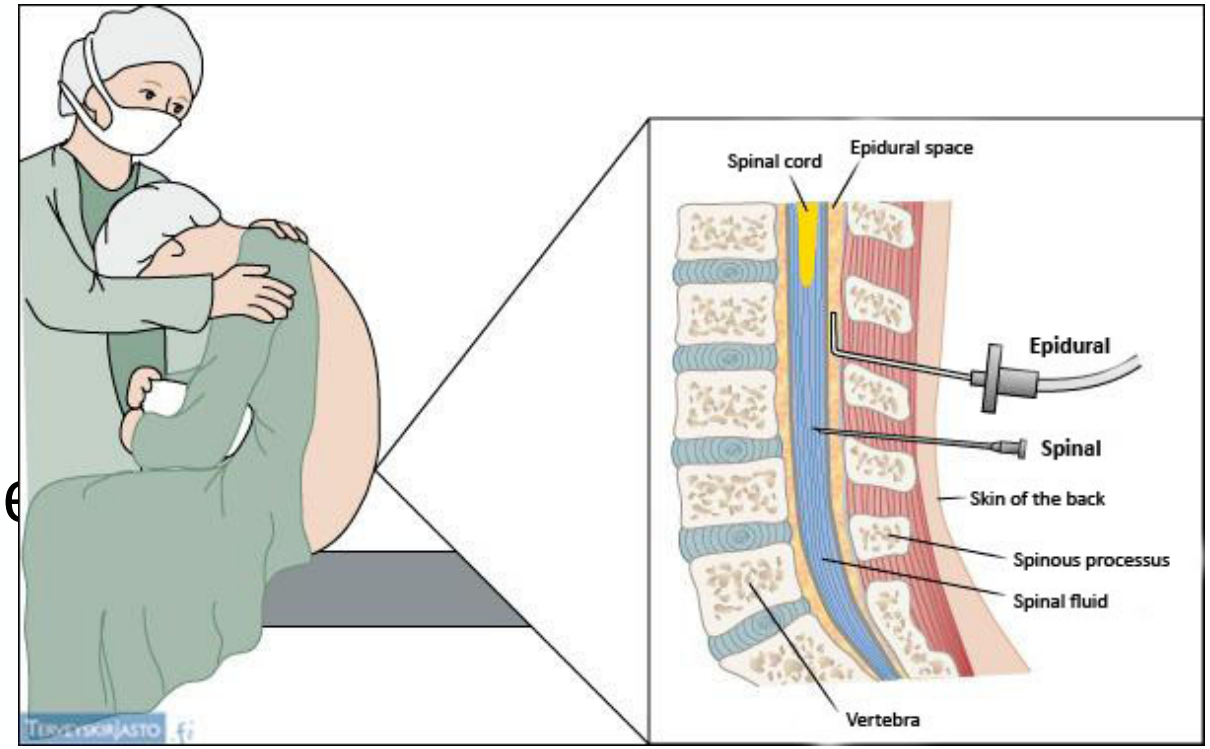
IVF/Surrogacy

Case Report Bergerson 2014

Patient had history of multiple arterial aneurysms, dissections. Had a splenic artery aneurysm rupture during stimulated cycle. Did numerous unstimulated cycles, unsuccessful implantation of embryo.

Obstetrics: Anesthesia

- Older studies state regional anesthesia contraindicated, but recent studies do not find complications or contraindications.
- Local anesthesia
  - May need higher dose and/or longer time to take effect.



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Obstetrics: Bleeding

- Significant bleed: 1,000 cc or more estimated blood loss, or required transfusion
- cEDS, vEDS associated with higher rates of bleeding
- Overall risk 19%
- Intrapartum bleeding
- Postpartum bleeding

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Other obstetrical complications

- Deep vein thrombosis 4%
- Coccyx dislocation

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Obstetrics: management

- Evaluate aortic root preconception
- Plan delivery at hospital with full services
- Monitor cervix 16-20 weeks gestational age
- Consult with anesthesia, especially if scoliosis, POTS, vEDS
- Use of DDAVP for control of hemorrhage, acts on platelets

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