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Improvement
Let's talk about "OHVIRA syndrome"
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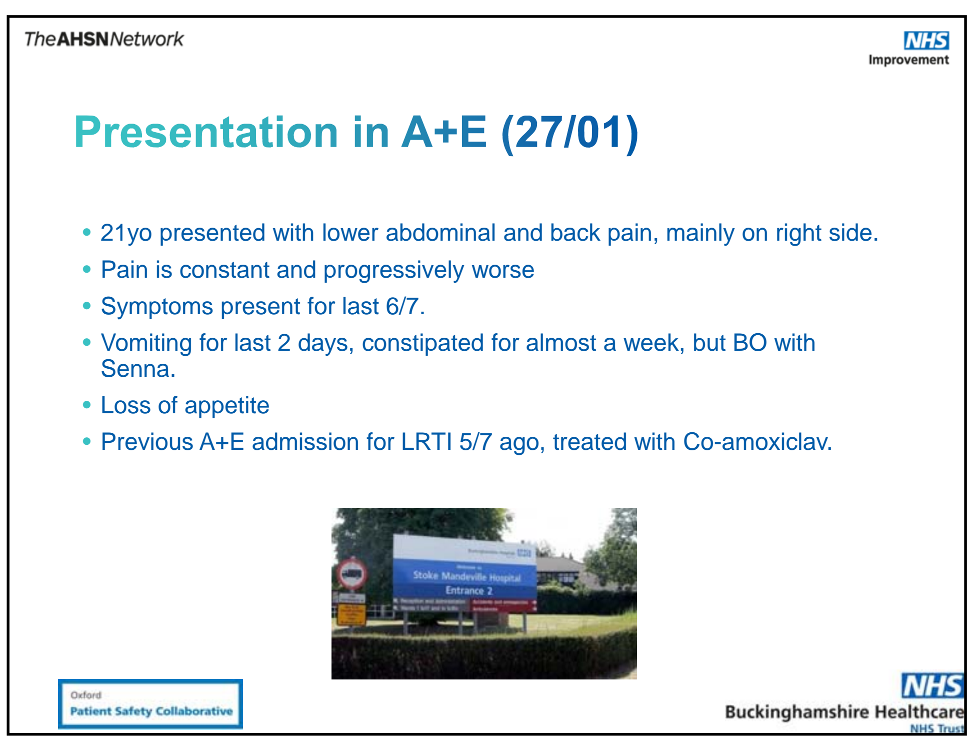
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Presentation in A+E (27/01)
21yo presented with lower abdominal and back pain, mainly on right side.
Pain is constant and progressively worse
Symptoms present for last 6/7.
Vomiting for last 2 days, constipated for almost a week, but BO with Senna.
Loss of appetite
Previous A+E admission for LRTI 5/7 ago, treated with Co-amoxiclav.
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Entrance 2
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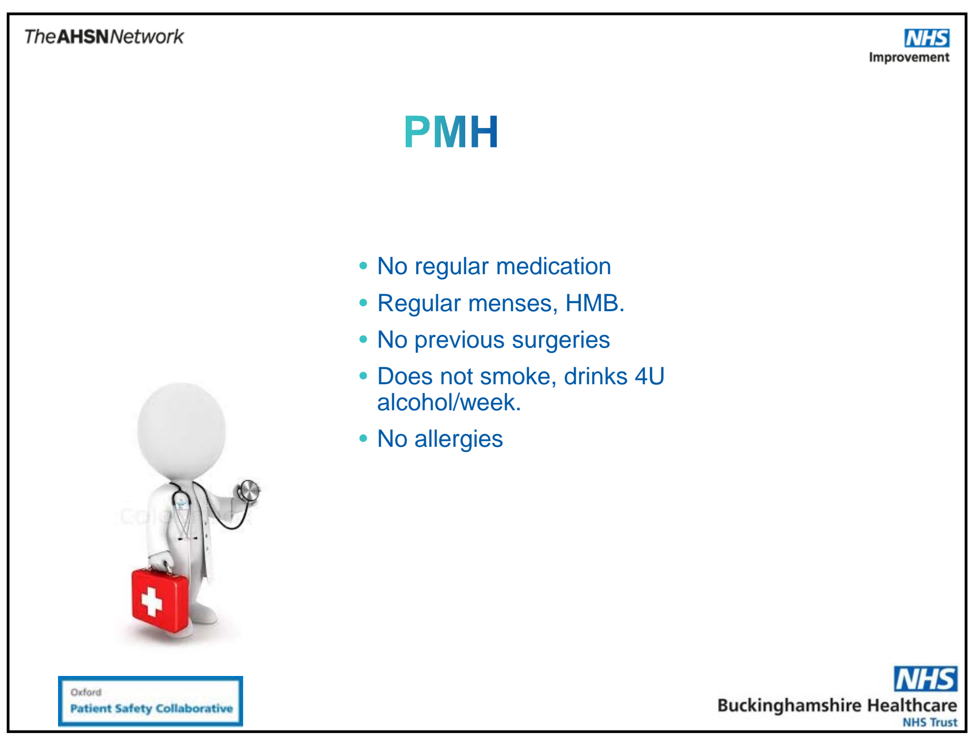
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PMH
No regular medication
Regular menses, HMB.
No previous surgeries
Does not smoke, drinks 4U alcohol/week.
No allergies
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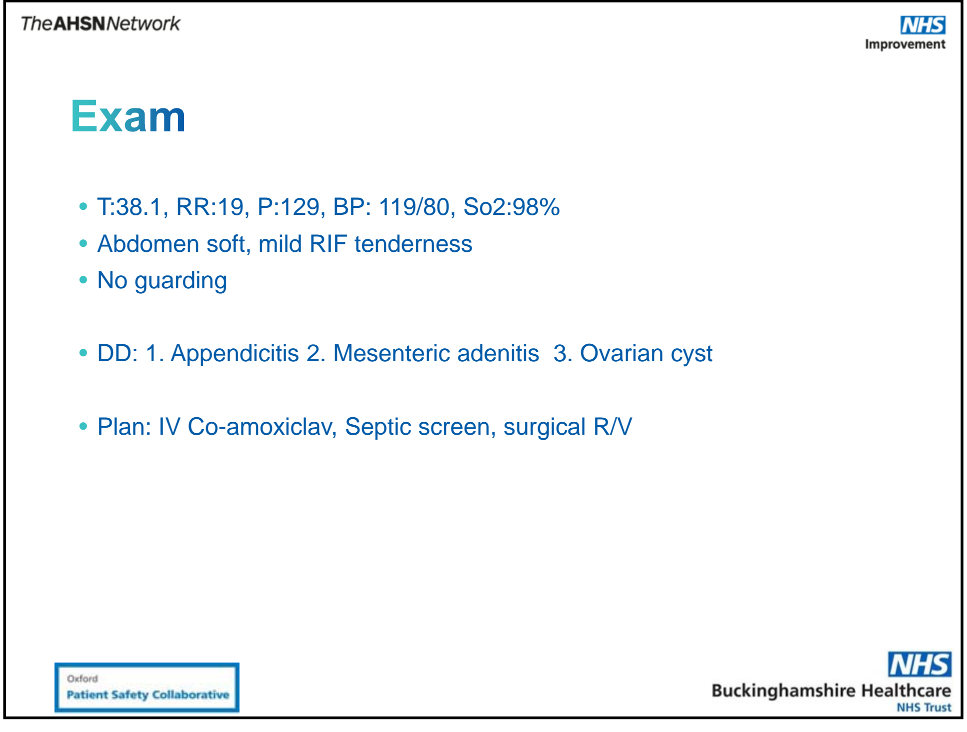
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Exam
T:38.1, RR:19, P:129, BP: 119/80, So2:98%
Abdomen soft, mild RIF tenderness
No guarding
DD: 1. Appendicitis 2. Mesenteric adenitis 3. Ovarian cyst
Plan: IV Co-amoxiclav, Septic screen, surgical R/V
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Investigations

- VBG: pH: 7.35, Lac: 1.3
- Urine: (+) leucocytes, protein, blood, 3(+) ketones, UPT negative
- Bloods: Hb:127, WCC: 16, CRP>320, ALT:36, U+Es: normal.
- CT: 1. Complex pelvic collection (7 x 6 x 10cm), arises from left adnexa, inflammatory process extending into the uterus. Suggestive of TOA 2. Right renal agenesis 3. Focal fatty liver infiltration
- Refer to Gynae

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Gynae R/V

- LMP: 2 weeks ago
- Regular periods with dysmenorrhea and HMB
- Regular sexual partner for last few years
- No issues with intercourse

- Speculum + VE: Cervix difficult to visualise, deviated to the left, purulent foul-smelling discharge, fullness felt on anterior fornix, no tenderness. Swabs sent
- Plan: 1. PID abs 2. USS 3. Chase swabs

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28/01

- Stills spiking (on Clindamycin + Gentamicin due to mild allergic reaction to initial regimen).
- Bloods: CRP: 288, WCC: 17.
- USS: 7 x 6.5 x 6.5 cm complex, cystic mass in RIF. Ovaries not clearly visualised. Uterus didelphys.

- Possibility of OHVIRA syndrome raised.
- Plan: For MRI

"Everything is going to be fine, Mrs. Witzer! An orderly is getting a can of Crisco™ and a winch, and we'll have you out of there soon!"

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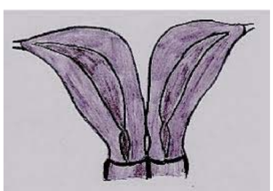
29/01

- Still spiking and tachycardic
- CRP: 286, WCC: 15, PLT: 498
- Metronidazole added to antibiotic regimen
- MRI: There is a double uterus and cervix. There is a longitudinal vaginal septum, and the right-sided hemivagina is obstructed, with evidence of haematocolpos. There is also a gas fluid level within its lumen, indicating that the fluid contents may be infected. Probable right hydrosalpinx/TOA noted.
- OHVIRA diagnosis confirmed.



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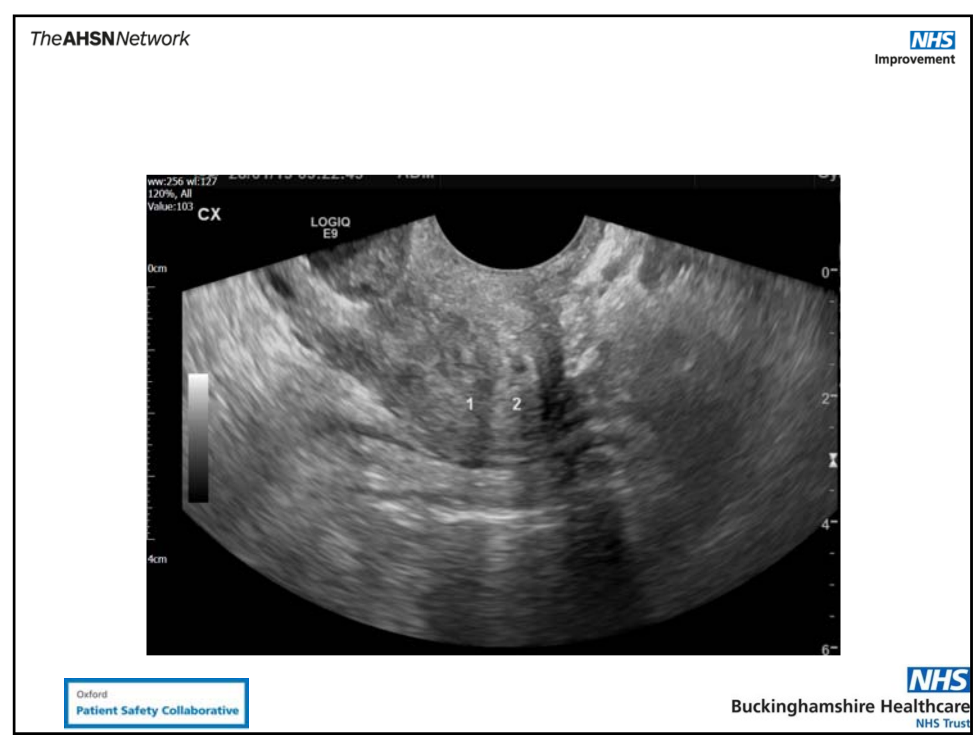
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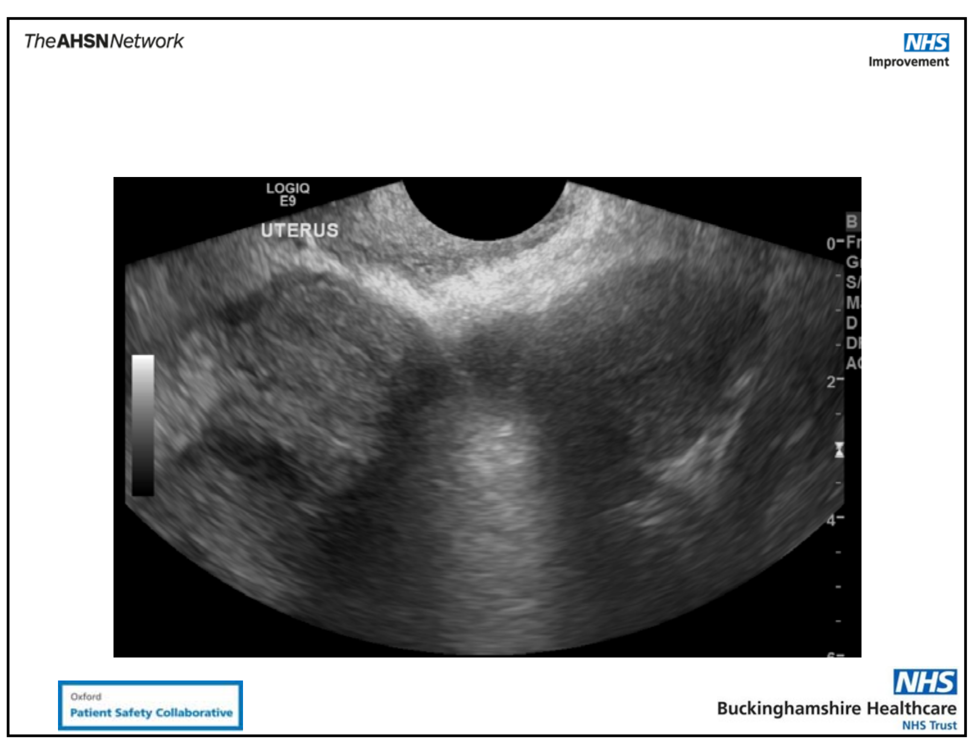
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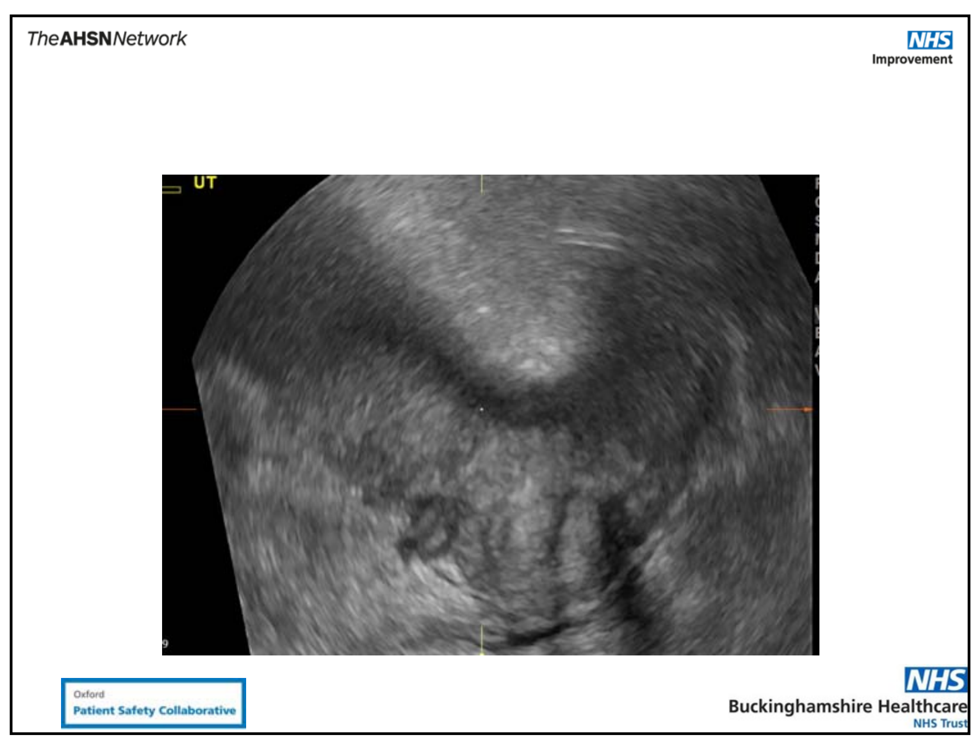
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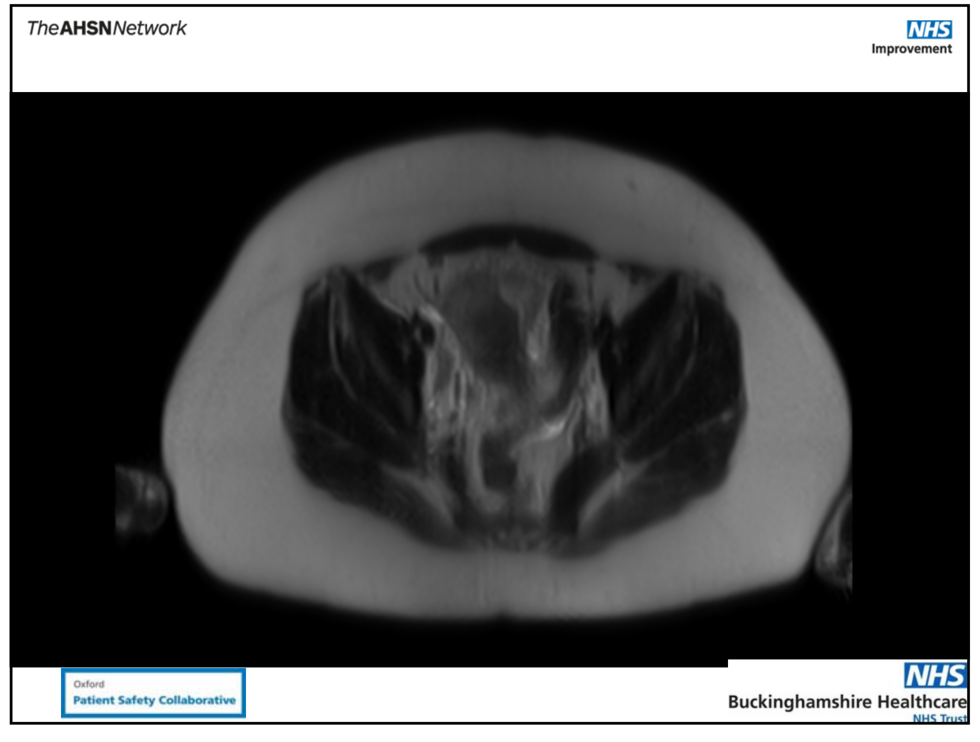
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30/01

- Still spiking
- CRP: 242, WCC: 16.5, PLT:500
- Plan for joint EUA + division of vaginal septum and drainage of haematocolpos +/- laparoscopic drainage of tubo-ovarian abscess on 01/04
- 31/01: Blood cultures negative, inflammatory markers static. Still spiking.

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01/04

- Vaginal septectomy and drainage of pyocolpos + laparoscopic adhesiolysis and drainage of tubo-ovarian abscess.
- Significant bowel adhesions and large left-sided abscess were noted.

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Post-operative recovery

- Inpatient until D4 post-op
- Abdominal drain in situ for 3 days
- Continued IV Meropenem + Doxycycline
- Stopped spiking temperature post-op, pulse normalised after 2 days.
- CRP dropped to 51 and WW to 9.5
- HVS: Mixed anaerobes, sensitive to Metronidazole
- ECS: negative
- Blood cultures negative
- Discharged with a follow up in 4 weeks

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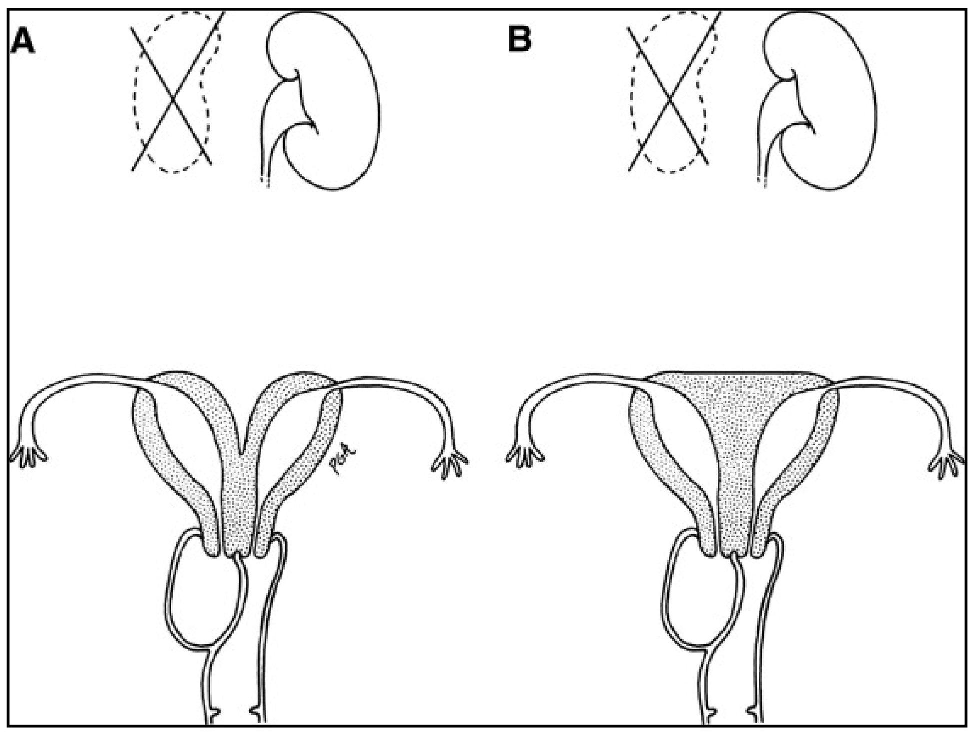
Literature review

- Also called Herlyn-Werner-Wunderlich syndrome
- Almost 300 cases reported
- Rare syndrome
- First described in 1922
- Typically associated with a didelphys uterus with two cervices and two vaginas, one of which is obstructed. The obstruction usually occurs on the same side as the renal anomaly.
- Renal anomaly is usually a renal agenesis, but also can be dysplastic kidney or duplex kidney.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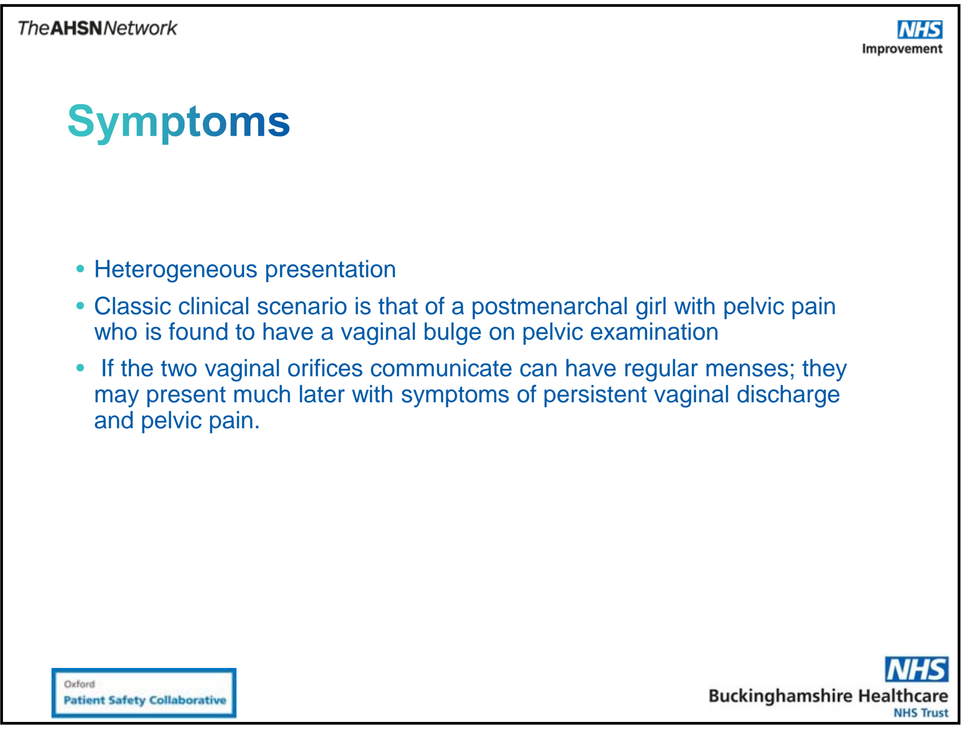
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Symptoms
• Heterogeneous presentation
• Classic clinical scenario is that of a postmenarchal girl with pelvic pain who is found to have a vaginal bulge on pelvic examination
• If the two vaginal orifices communicate can have regular menses; they may present much later with symptoms of persistent vaginal discharge and pelvic pain.
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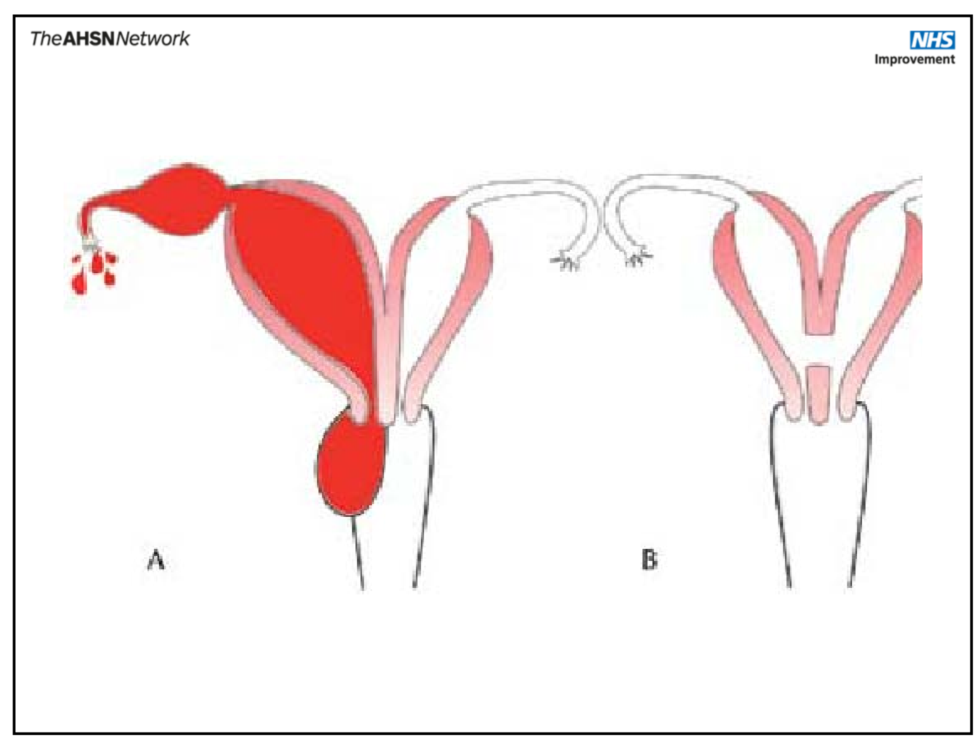
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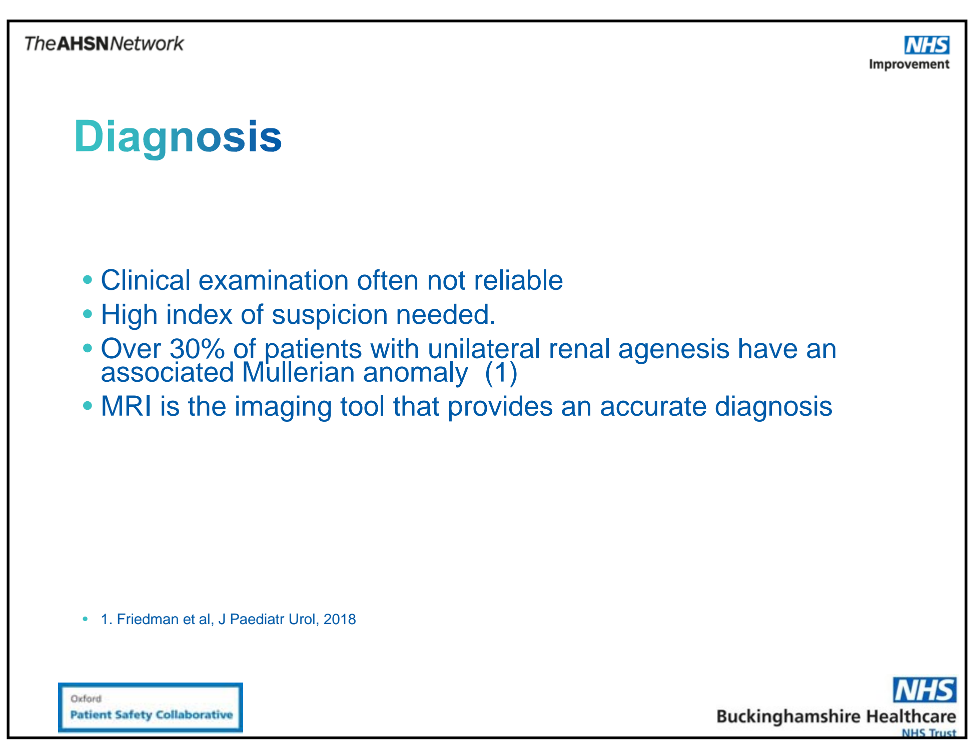
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Diagnosis
• Clinical examination often not reliable
• High index of suspicion needed.
• Over 30% of patients with unilateral renal agenesis have an associated Mullerian anomaly (1)
• MRI is the imaging tool that provides an accurate diagnosis
• 1. Friedman et al, J Paediatr Urol, 2018
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Treatment

- Excision of the obstructing septum followed by a marsupialization of the blind hemivagina is generally the treatment of choice.
- In all patients pain symptoms resolve. Dysmenorrhea resolves in 87% of cases.
- Dyspareunia also resolves if present before diagnosis.
- Laparoscopic drainage of TOA can be performed.
- Some advocate a two-stage procedure: the first surgery to reduce the hematocolpos, and the second to resect the excess septum after a period of wound-healing and vaginal remodeling.
- Hemihysterectomy and ipsilateral hemicolpectomy have been reported in extreme cases.

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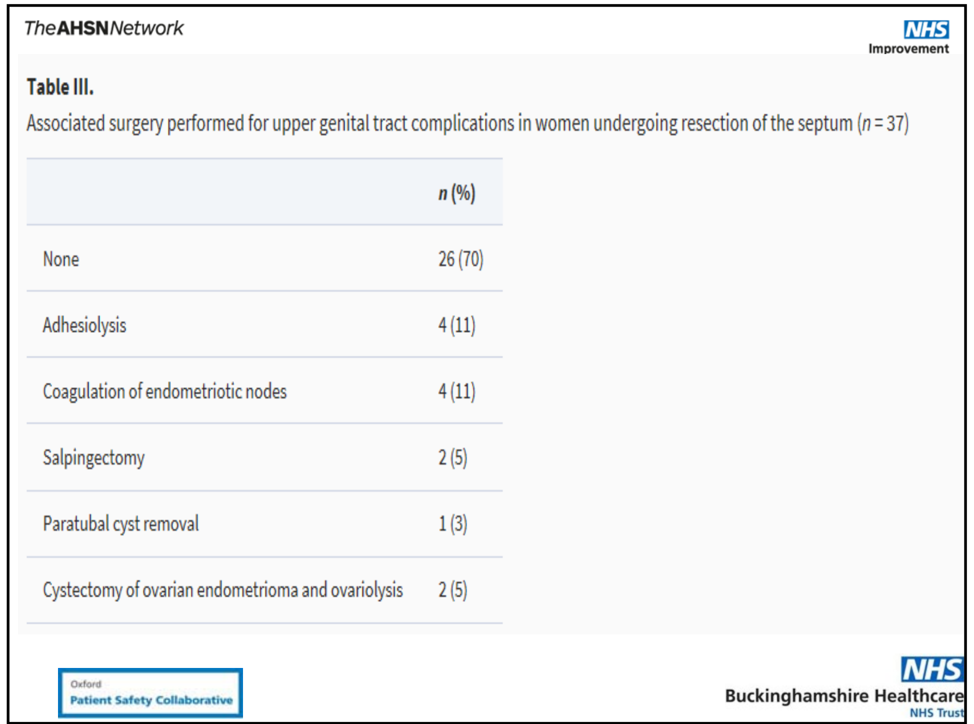

Table III.

Associated surgery performed for upper genital tract complications in women undergoing resection of the septum ($n = 37$)

	n (%)
None	26 (70)
Adhesiolysis	4 (11)
Coagulation of endometriotic nodes	4 (11)
Salpingectomy	2 (5)
Paratubal cyst removal	1 (3)
Cystectomy of ovarian endometrioma and ovariolysis	2 (5)

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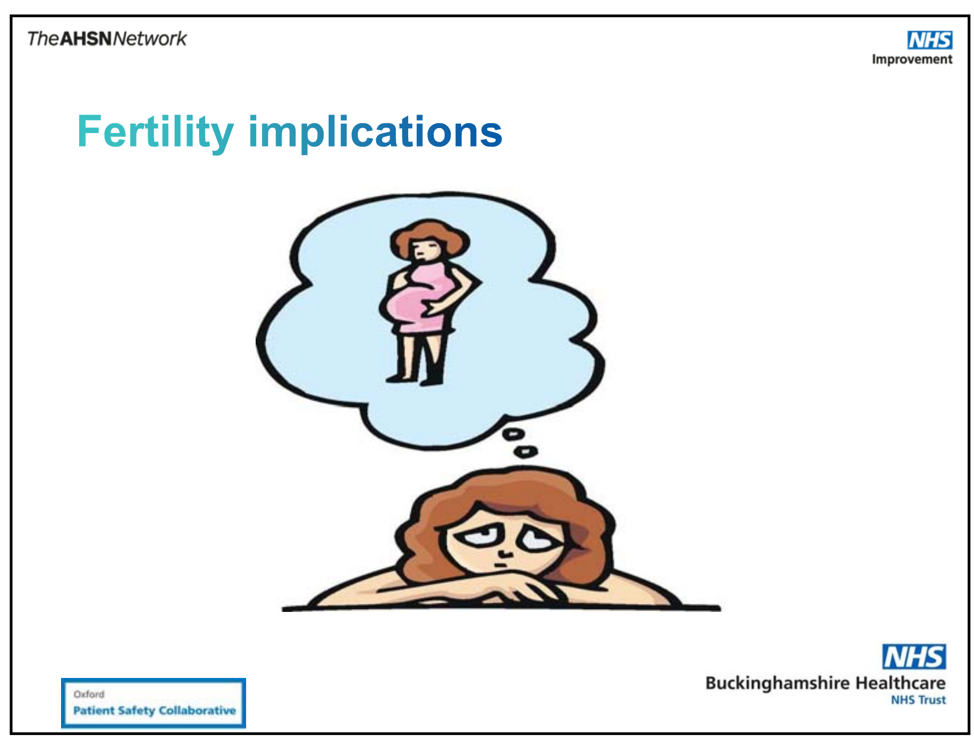
Fertility implications



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Blind hemivagina: long-term follow-up and reproductive performance in 42 cases FREE

B. Haddad, E. Barranger, B.J. Paniel

Human Reproduction, Volume 14, Issue 8, 1 August 1999, Pages 1962–1964,
https://doi.org/10.1093/humrep/14.8.1962
Published: 01 August 1999 **Article history ▾**

- 78% had a didelphic uterus and 22% a complete septated uterus
- 27 out of 42 did not wish to become pregnant
- Nine women who had undergone vaginal septum excision experienced 20 pregnancies after surgical procedure
- Pregnancies occurred mainly in the contralateral cavity (80%)
- Four out of 7 women with didelphic uterus had a preterm birth
- The five patients who underwent hemihysterectomy and ipsilateral hemicolpectomy did not achieve pregnancy.

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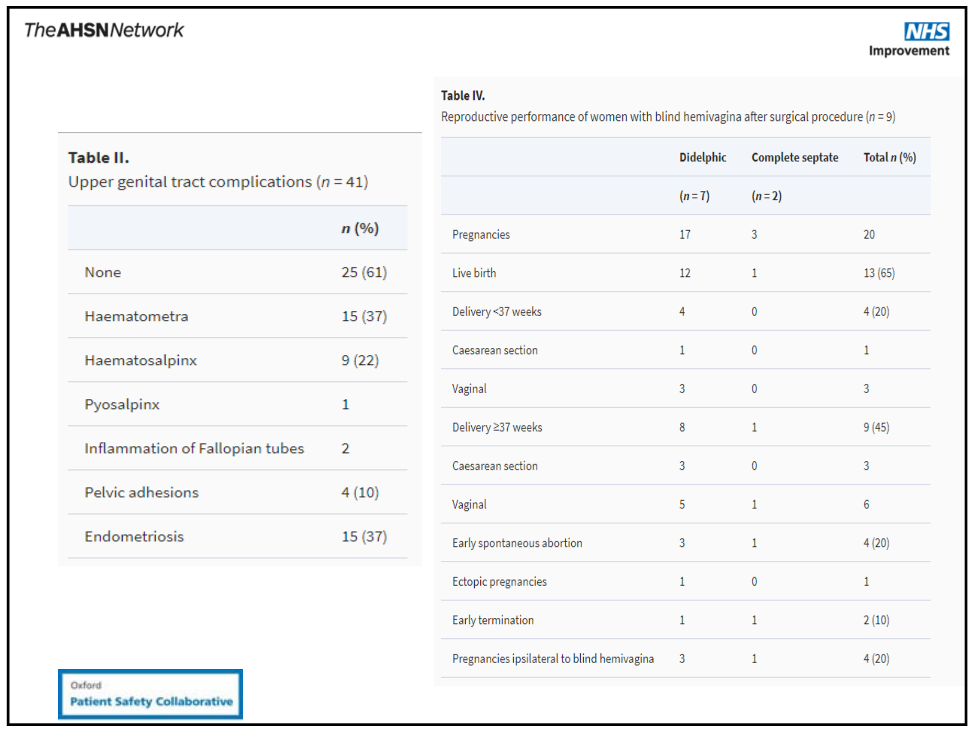
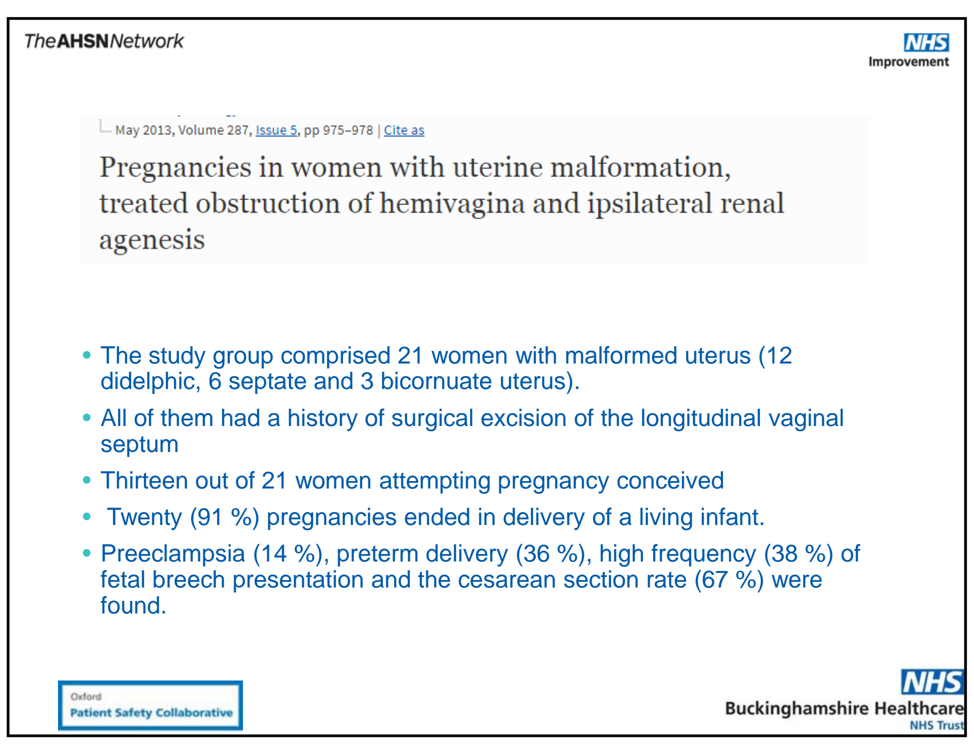


Table II.
Upper genital tract complications ($n = 41$)

	n (%)
None	25 (61)
Haematometra	15 (37)
Haematosalpinx	9 (22)
Pyosalpinx	1
Inflammation of Fallopian tubes	2
Pelvic adhesions	4 (10)
Endometriosis	15 (37)

Table IV.
Reproductive performance of women with blind hemivagina after surgical procedure ($n = 9$)

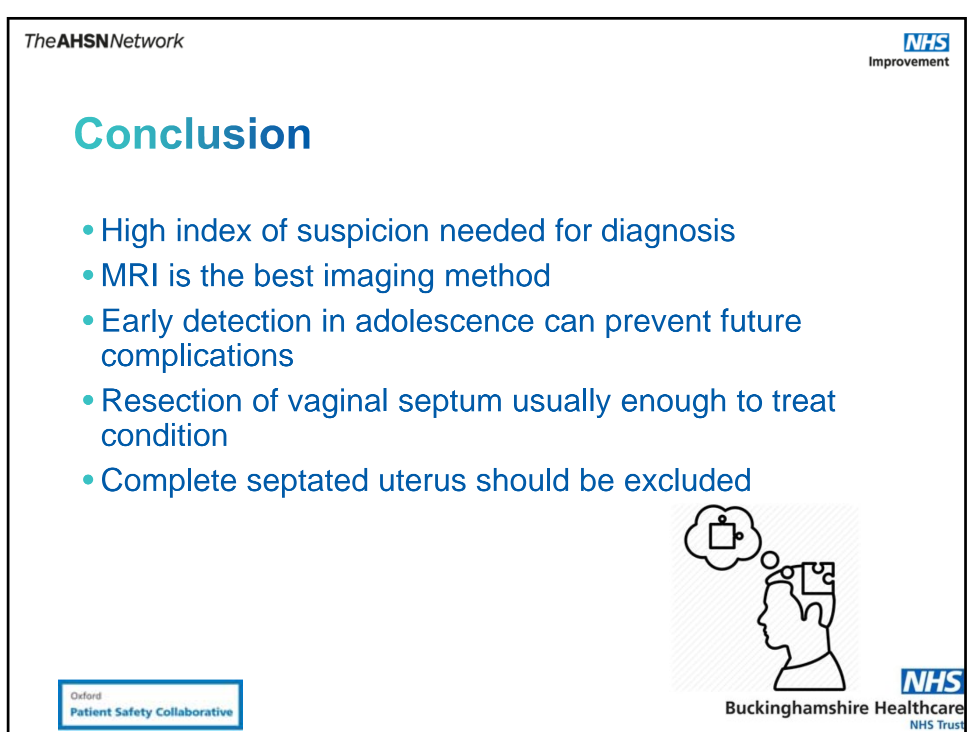
	Didelphic	Complete septate	Total n (%)
	($n = 7$)	($n = 2$)	
Pregnancies	17	3	20
Live birth	12	1	13 (65)
Delivery <37 weeks	4	0	4 (20)
Caesarean section	1	0	1
Vaginal	3	0	3
Delivery ≥37 weeks	8	1	9 (45)
Caesarean section	3	0	3
Vaginal	5	1	6
Early spontaneous abortion	3	1	4 (20)
Ectopic pregnancies	1	0	1
Early termination	1	1	2 (10)
Pregnancies ipsilateral to blind hemivagina	3	1	4 (20)



May 2013, Volume 287, Issue 5, pp 975–978 | Cite as

Pregnancies in women with uterine malformation, treated obstruction of hemivagina and ipsilateral renal agenesis

- The study group comprised 21 women with malformed uterus (12 didelphic, 6 septate and 3 bicornuate uterus).
- All of them had a history of surgical excision of the longitudinal vaginal septum
- Thirteen out of 21 women attempting pregnancy conceived
- Twenty (91 %) pregnancies ended in delivery of a living infant.
- Preeclampsia (14 %), preterm delivery (36 %), high frequency (38 %) of fetal breech presentation and the cesarean section rate (67 %) were found.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Conclusion
High index of suspicion needed for diagnosis
MRI is the best imaging method
Early detection in adolescence can prevent future complications
Resection of vaginal septum usually enough to treat condition
Complete septated uterus should be excluded
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Thank you
Mullerian Duct Anomalies
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Special thanks
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•Mr Warakaulle, Consultant Radiologist
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