

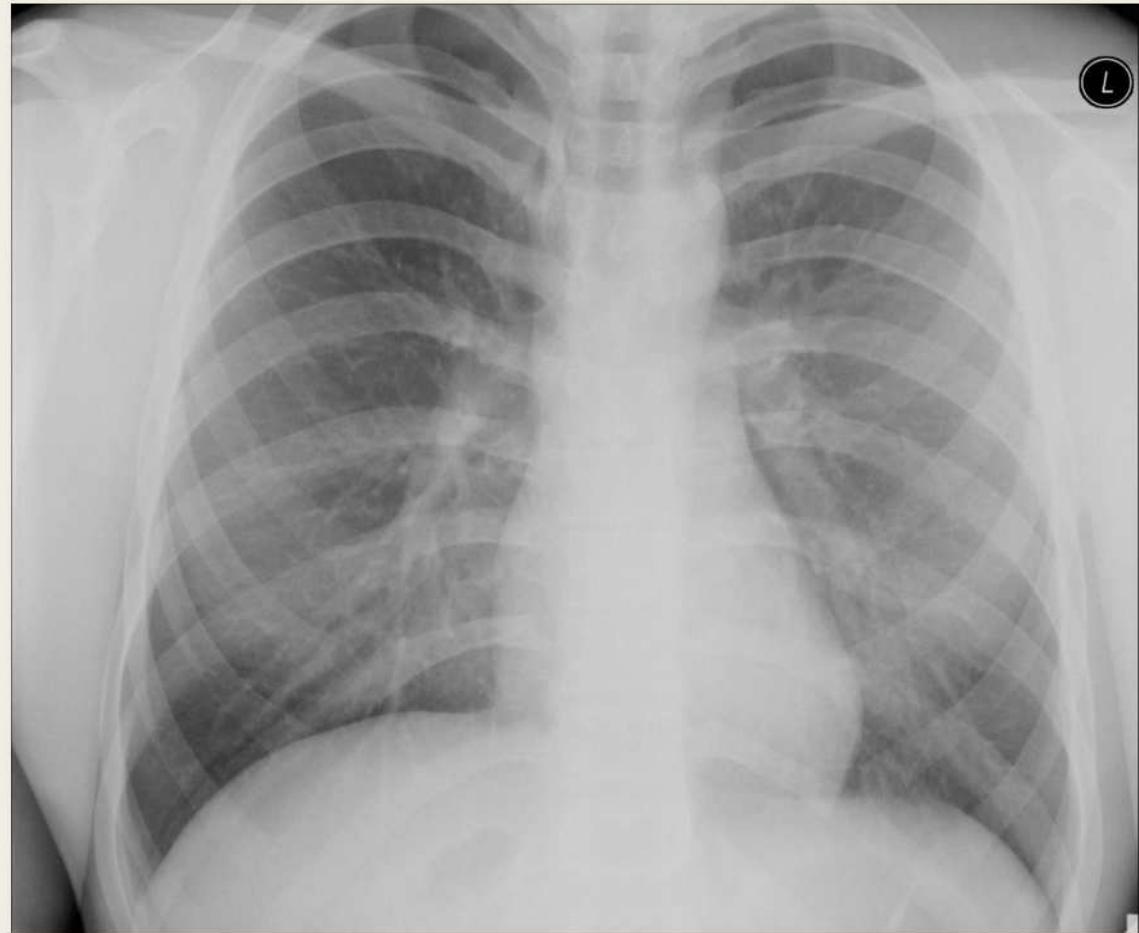
HAIGUSJUHT

23 AASTANE MEES ALAKÕHUVALUGA

Jevgeni Kulikov
3. aasta resident

Anamnees

- 10.10.2017 öösel kõhuvalu, korduvalt oksendanud, oksemassid tavapärased, köht käib läbi.
- Kõhuvalu kirjeldab rohkem kui "vaev" alakõhus, mis surub põiele.
- Kaasuvaid haigusi pole teada
- Varasemad uuringud: Thorax rtg 2013, liigeste rtg.



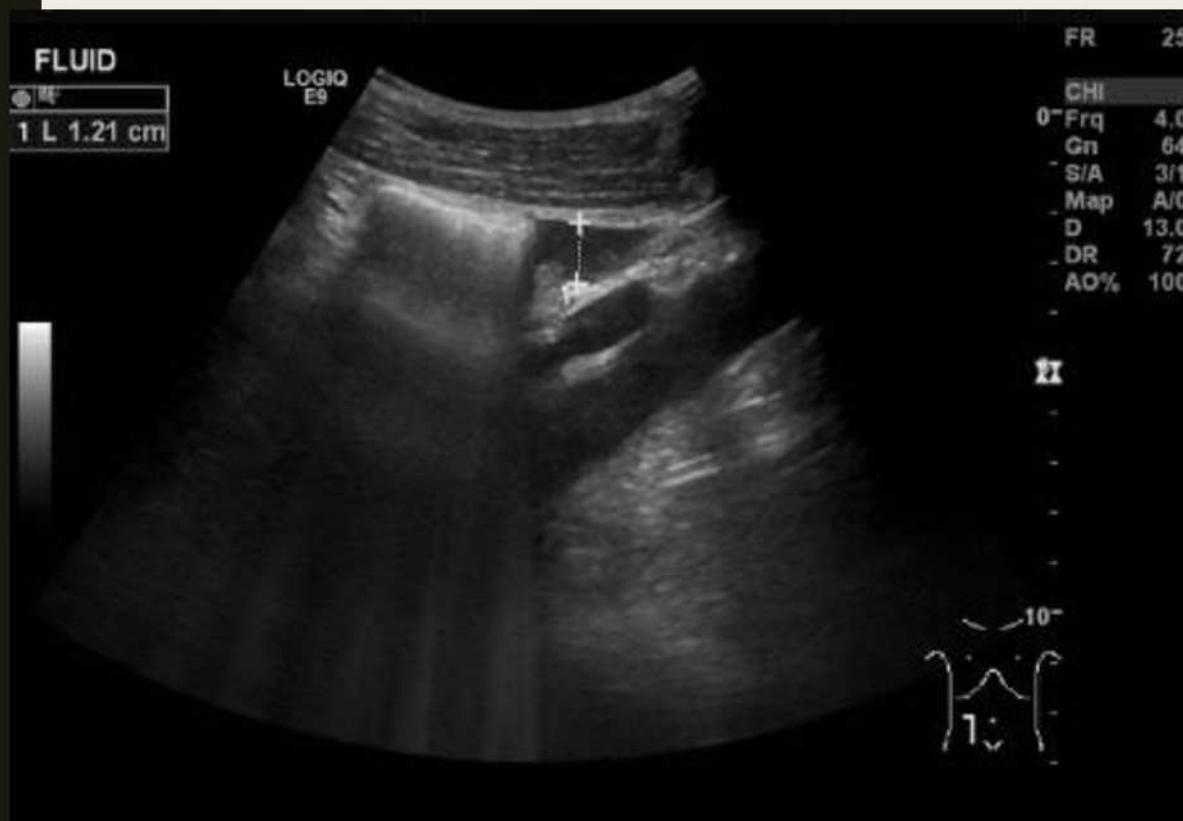
Objektiivne leid

- Obj. heas üldseisundis, kõhukatted pinges, suurim valu vasakul alakõhus. Blumberg alakõhus positiivne, enim vasakul. Rovsing-, Psoas -. Epigastrium piirkond valutu. Peristaltika tavapärane, tasane.
- Kehatemp. 36.3, WBC 12,3, neut 10,1 (83%), CRP <5 mg/L, ALAT/ASAT/GT/Bil/Amyl normis, uriin puhas
- Alvarado skoor – 7/10 (58-88% chance of positive appendicitis)

Tellitud UH

■Appenditsiit? Divertikuliit?

- Alakõhus piiratud nähtavus, sooled on promineeruva laiusega ja elavnenud peristaltikaga, **veenvat appendiksit** nende foonil **visualiseerida ei õnnestu**.
Alakõhus soolte vahel on kohati **vähest vaba vedelikku**.



- Maks on laterolateraases suunas suurenenud, ulatub vasaku neeru ülapooluseni, põrna visualiseerida ei õnnestu.
- Parema neeru ülapooluse ja maksa vahel lisastruktuurid - lümfisõlmed? migrerunud lisapõrnad?
- Soovitan täiendavana KT uuringut.

FR 18

0-CHI
- Frq 4.0
- Gn 64
- S/A 3/1
- Map A/0
- D 19.0
- DR 72
5-AO% 100

LOGIQ
E9

1 L 12.19 cm

LOGIQ
E9

1 L 7.13 cm

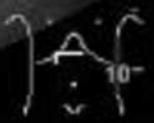
2 L 3.34 cm

FR 26

CHI
0-Frq 4.0
Gn 64
S/A 3/1
Map A/0
D 11.0
DR 72
AO% 100

II

5°



10°



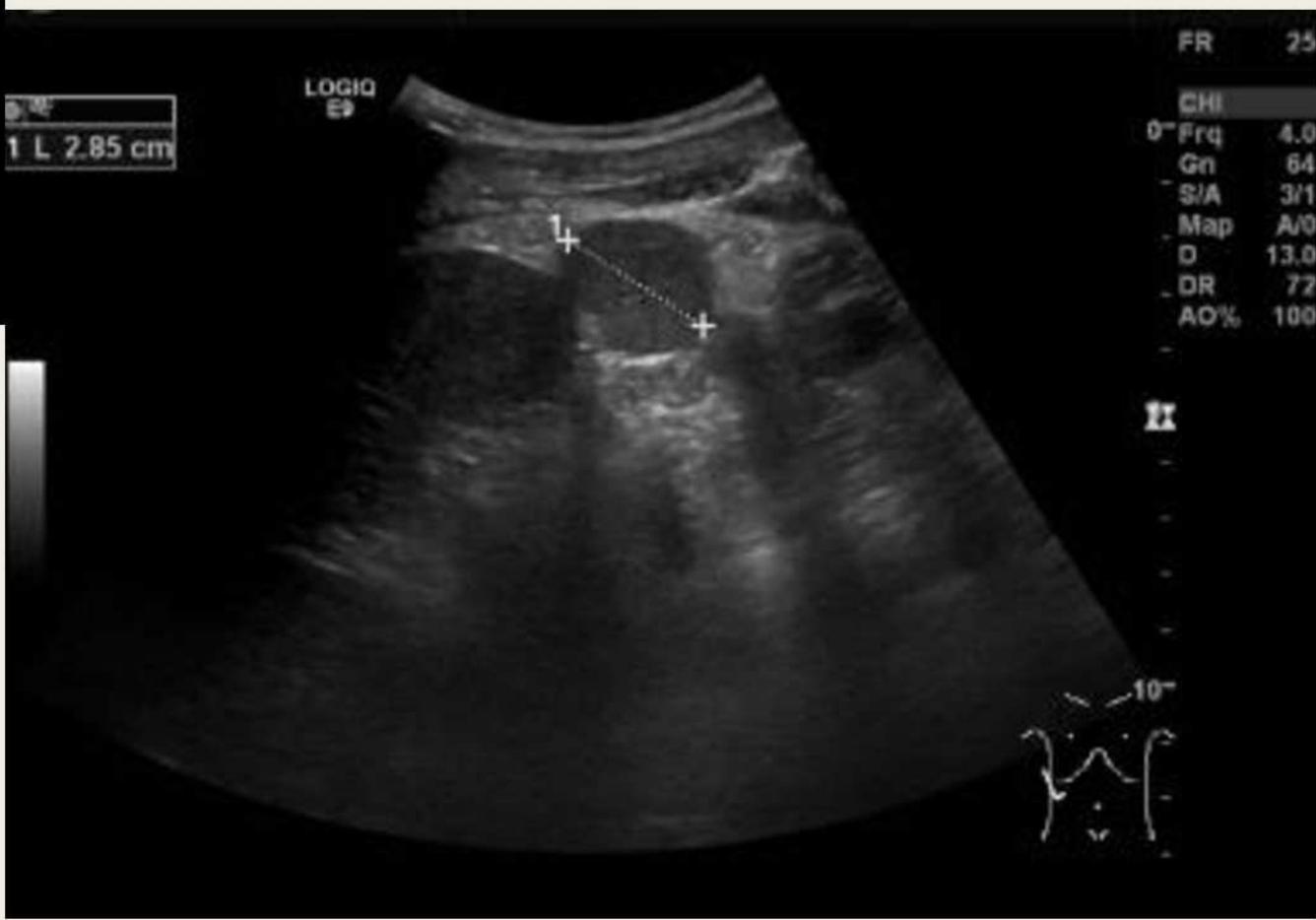
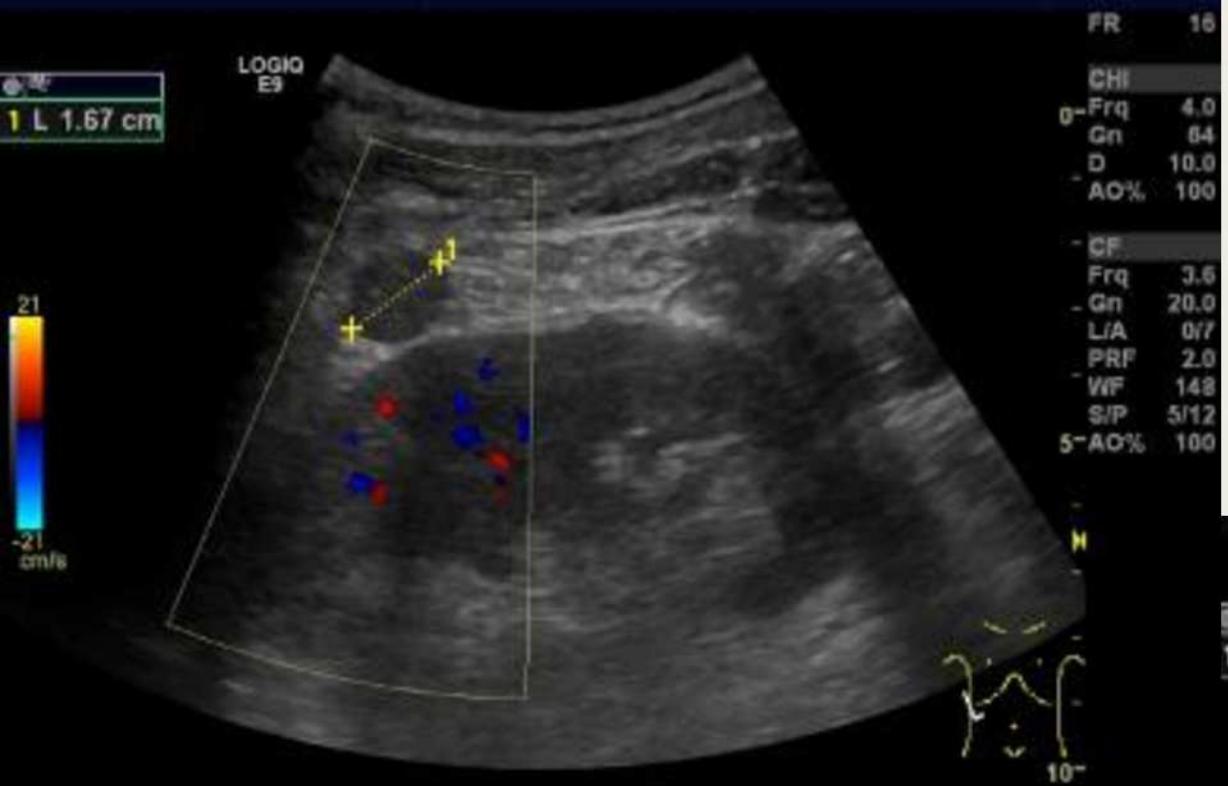
15°

S/A 3/1
Map A/0
D 12.0
DR 72
AO% 100

5°

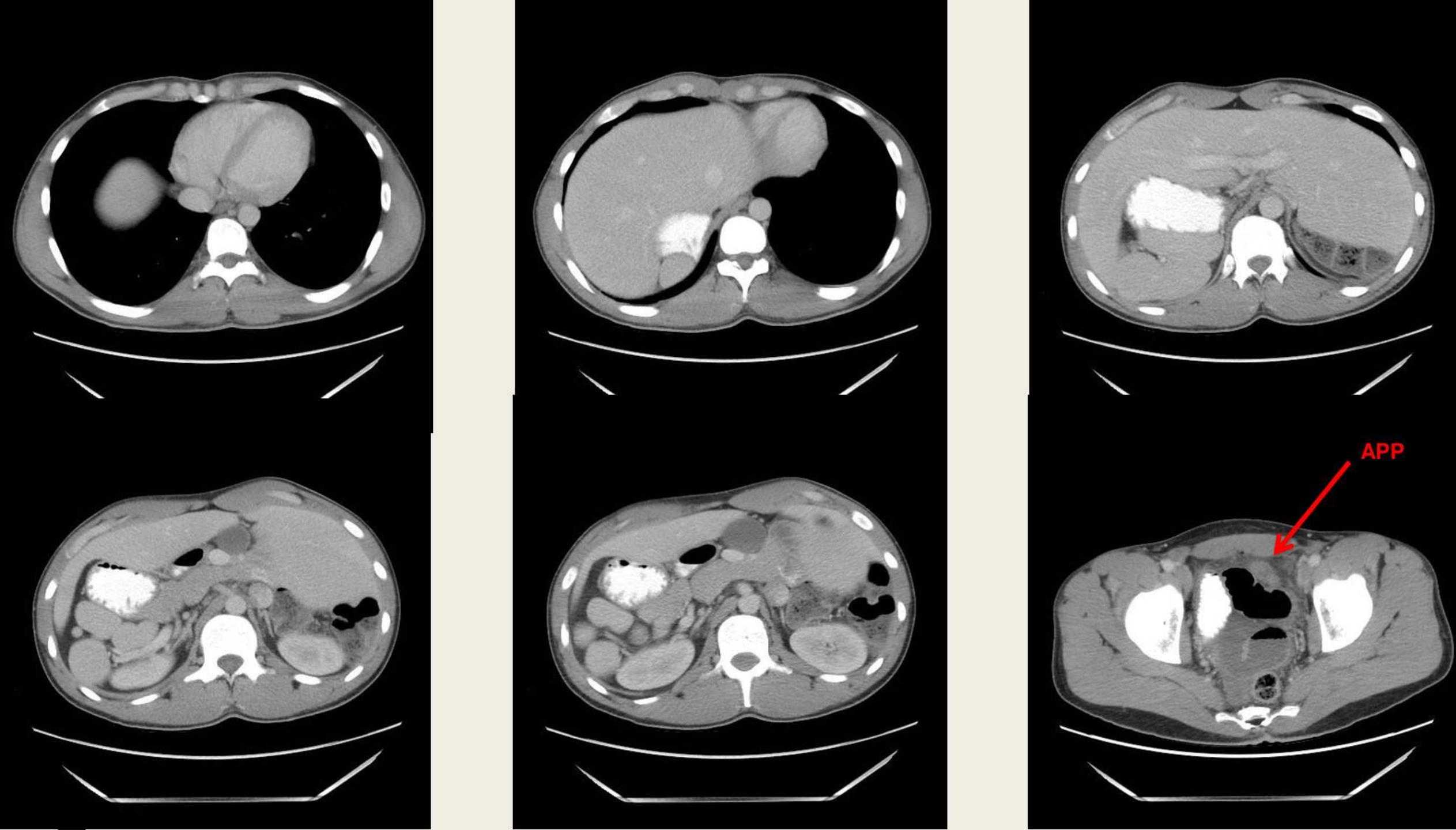


10°

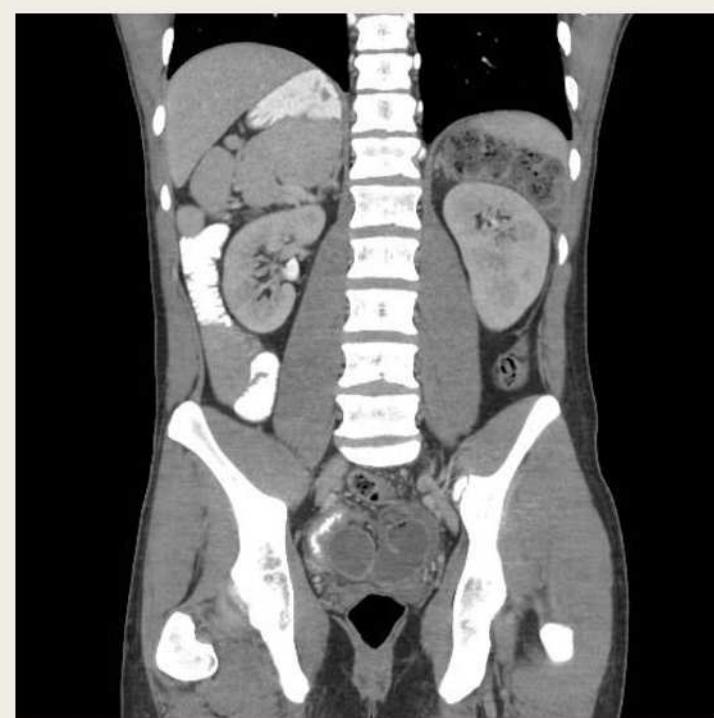
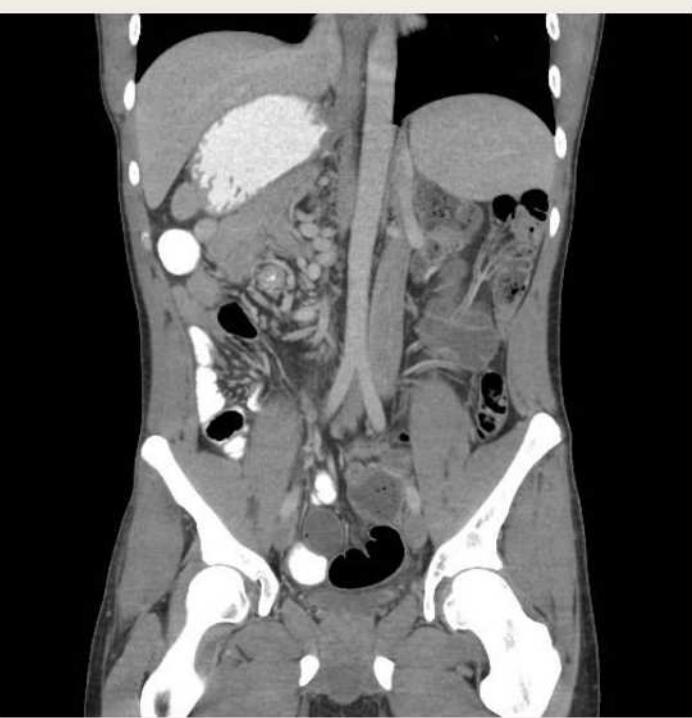
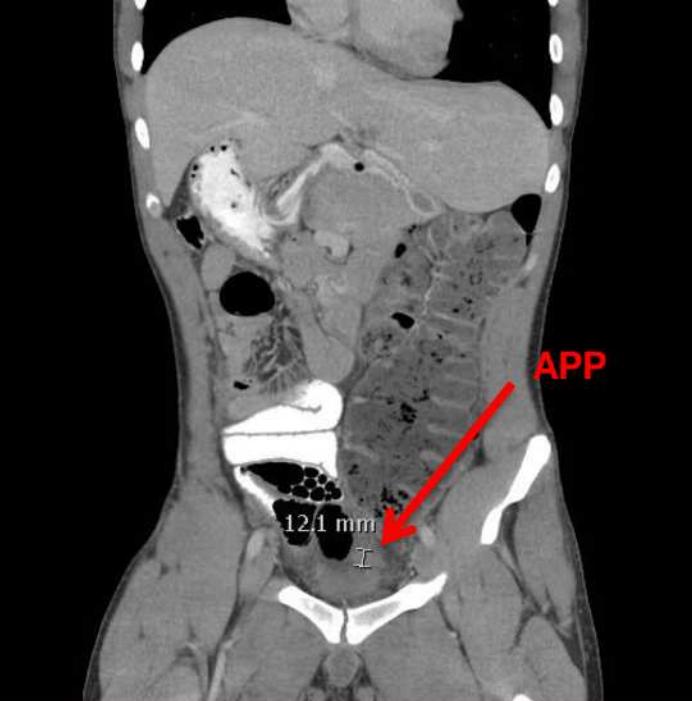


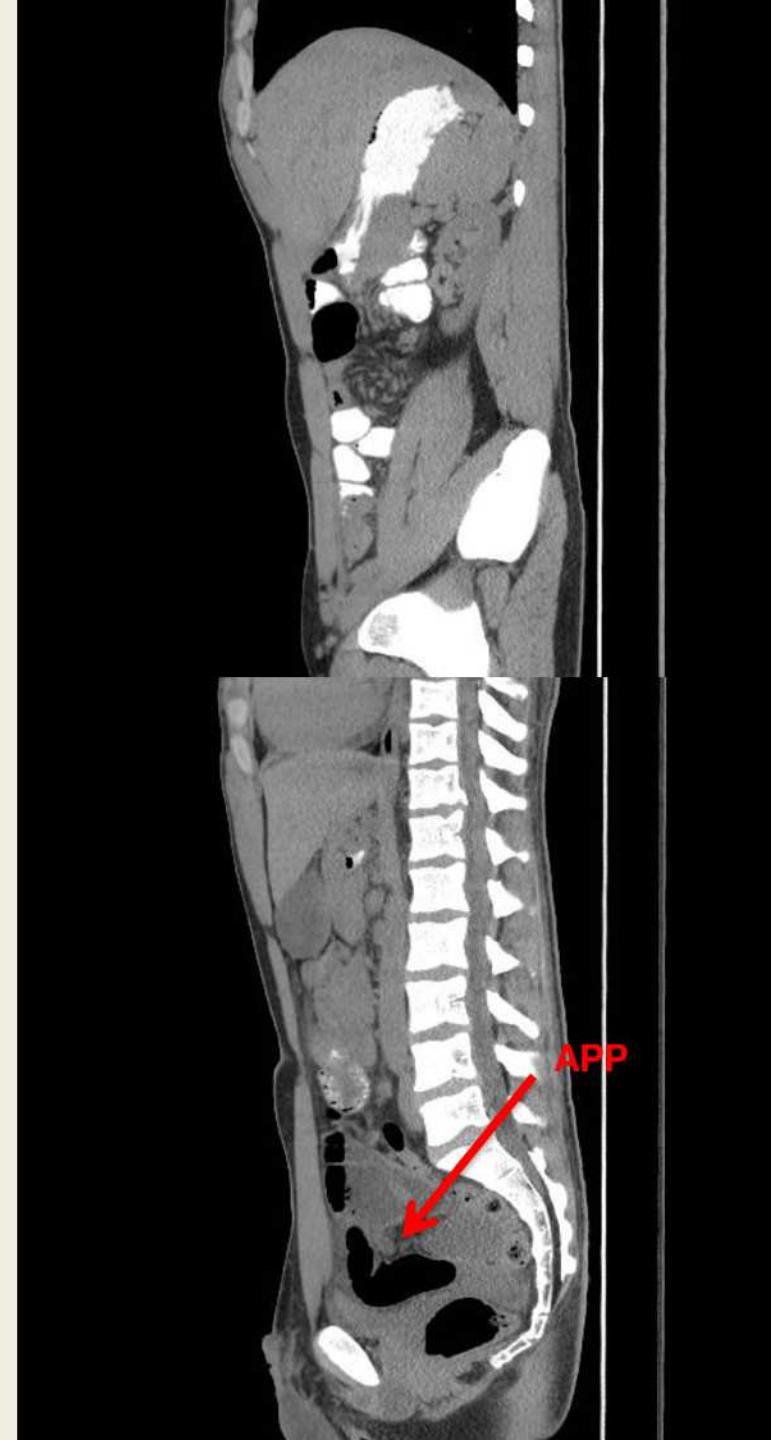
KT





APP





KT leid

Kõhuõõne organite asendianomaalia (**situs ambiguous polüspleeniaga**)

- Maks ja sapipõis keskjoonel
- Polysplenia (> 10) paremal
- Magu ja peensool paremal
- Pankrease pea vasakul
- IVC vasakul
- Kogu jämesool vasakul, laienenud
- **Põletikuline appendiks vasakul alakõhus**
- Vähene vaba vedelik

Operatsioon

- Laparoskoopiline apendektoomia. Troakaarid sisestatud paremalt poolt. Esineb soolte malrotatsioon - magu ja peensool on paremal pool kõhus ja suurem osa jämesoolest on vasakul pool kõhus. **Apendiks on vasakul pool** kõhus peen- ja jämesoole ülemineku kohal, **flegmonoosses põletikus**. Ei esine normaalset vaba rasvikut. **Kõhus on purulentne peritonit.**

Postop

- Postoperatiivne kulg soodne, alates 13.10 põletikunäitajate osas selge positiivne dünaamika ja patsient lubatud kodusele ravile heas üldseisundis.

SITUS ANOMAALIAD

Situs solitus

**Situs ambiguous
(Heterotaxy)**

Polysplenia



Ladina keelest:

"situs" - positsioon, lokalisatsioon

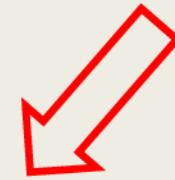
"solitus" - tavaline

"inversus" - tagurpidi, ümberpööratud

"ambiguous" - ebamääärane, ebakindel, varieeruv

Situs inversus

*Situs inversus
totalis
(Dextrocardiaga)*



*Situs inversus
incompletus
(Levocardiaga)*

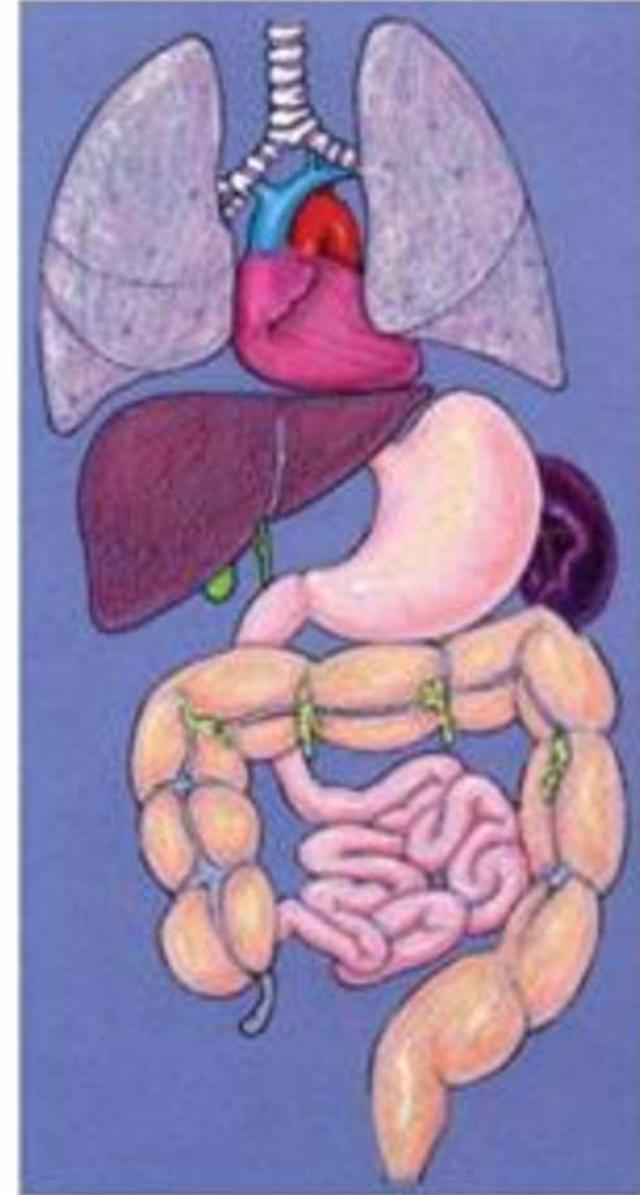


Situs solitus

„Situs solitus“ – normaalne organite positsioon

- parem kops 3 sagaraga, vasak 2 sagaraga
- südame apex, põrn, magu, aorta vasakul
- maks, VC paremal

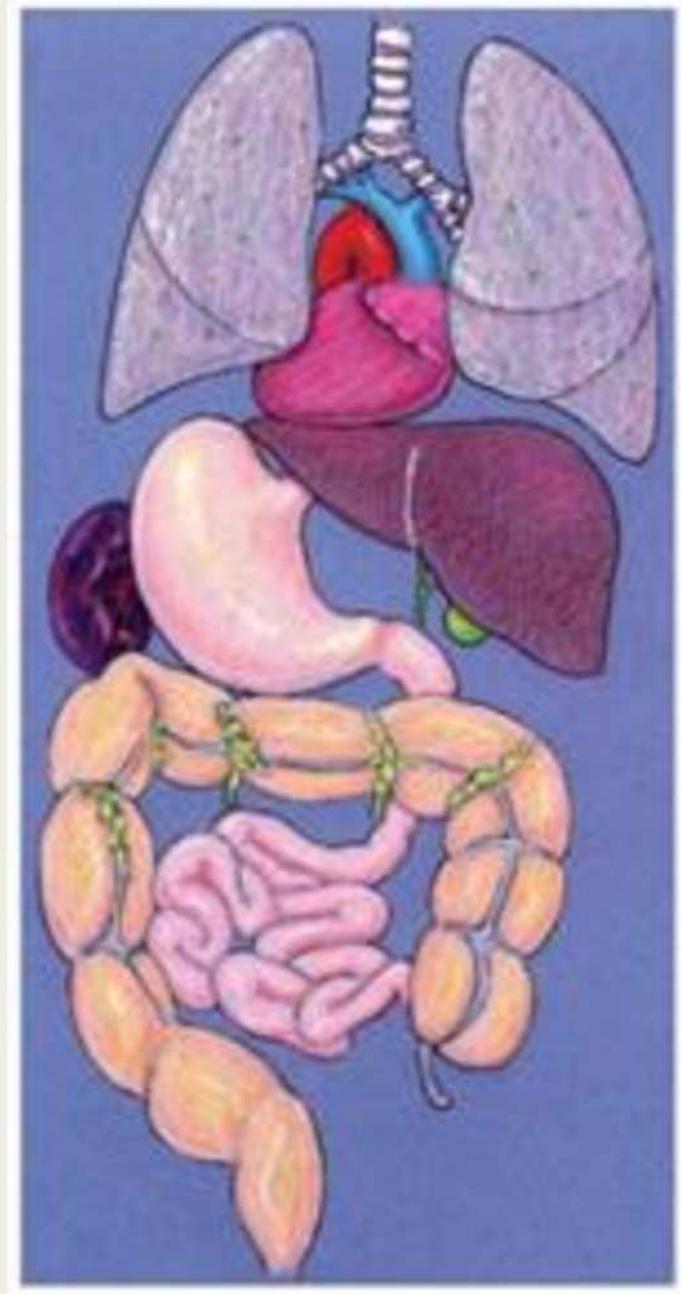
Kaasasündinud südamerikked 0,6-0,8%



Ann S. Fulcher "Abdominal manifestations of situs anomalies in adults" Radiographics 2002;

Situs inversus

- Normist peegelpilt
- Dextrocardiaga (totalis) ~0,01%
- Levocardiaga (incompletus) ~ 0.00005% (200 korda haruldasem)
- Kaasasündinud südamerikked:
 - 3-5% dextrocardiaga,
 - ~100% levocardiaga
- Seos Kartageneri sündroomiga



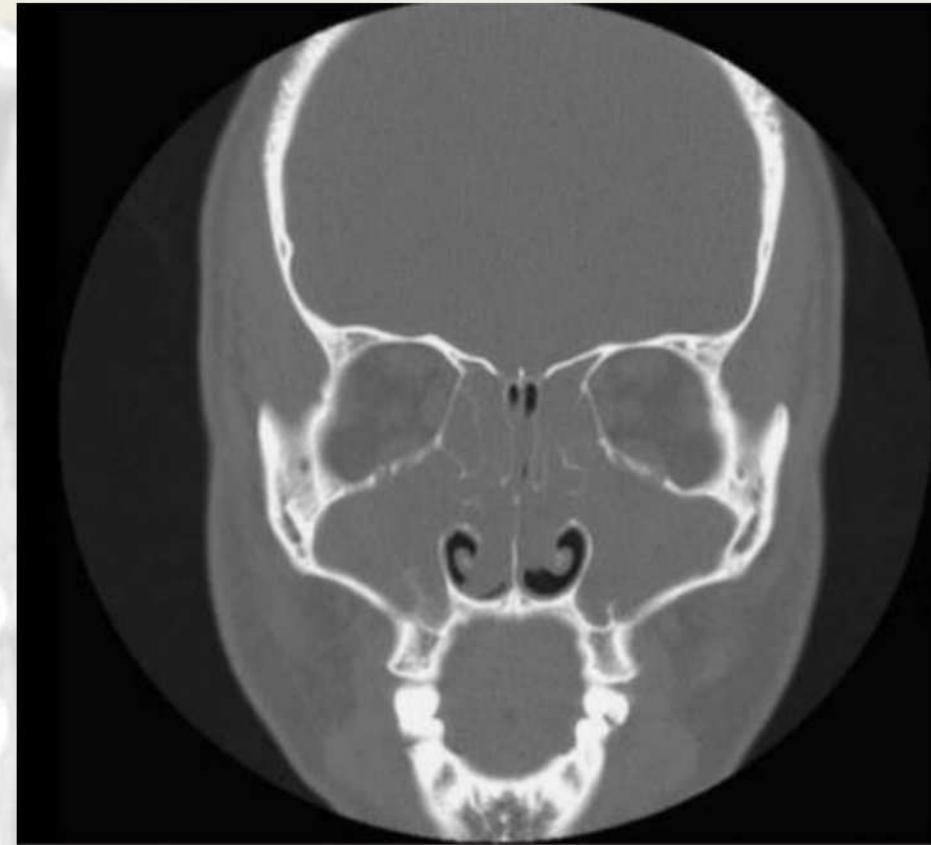
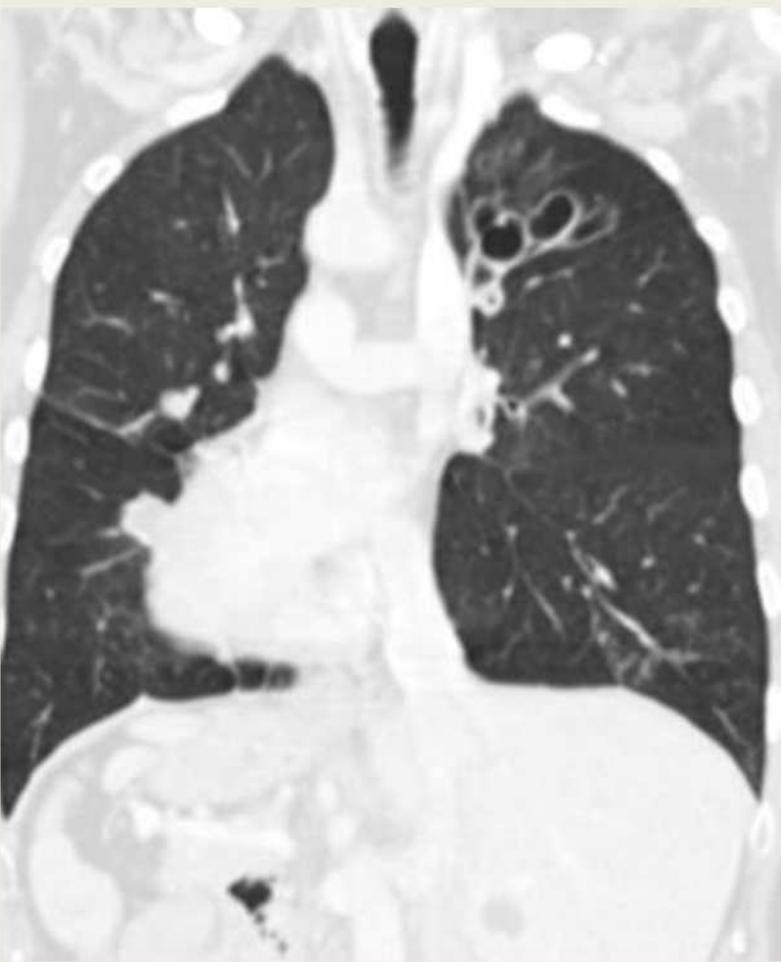
Ann S. Fulcher“Abdominal manifestations of situs anomalies in adults” Radiographics 2002;

Kartagener syndrome – primaarne tsiliaarne düskineesia

Triaad:

- 1) Situs inversus
- 2) Sinonasaalne polüpoos/kroonilin e sinusiit
- 3) Bronhektaasiad

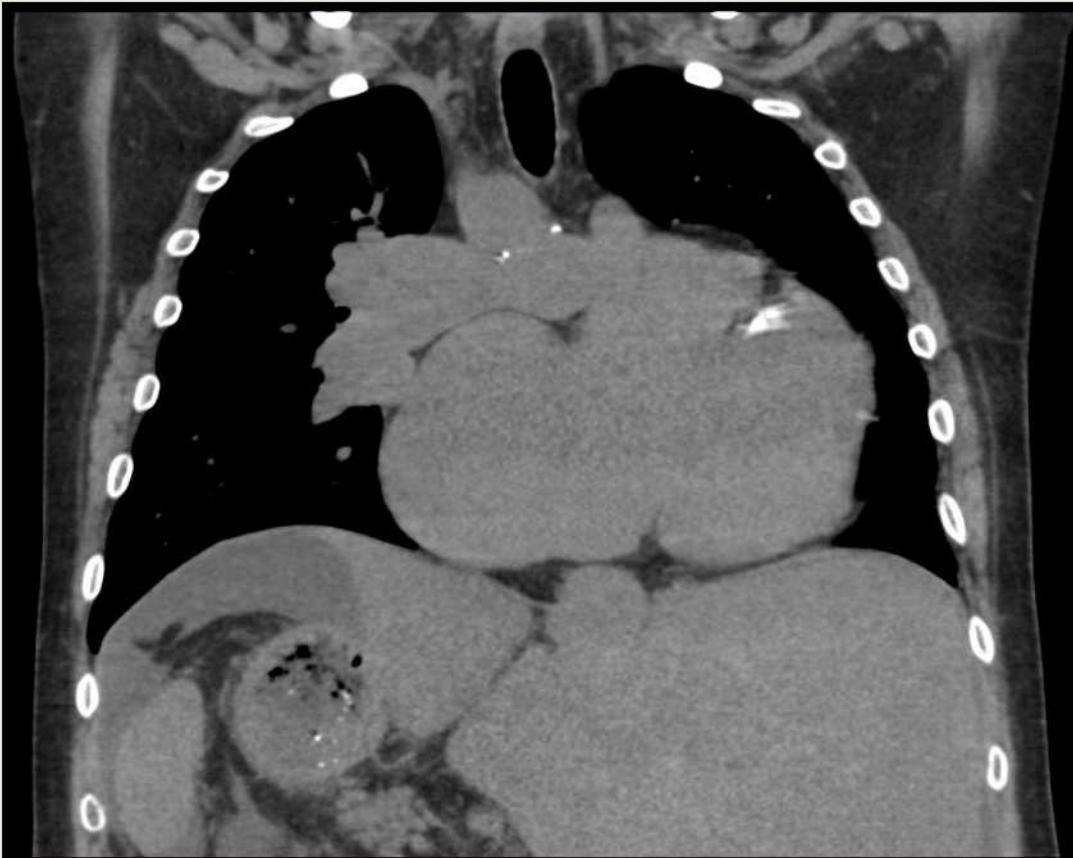
~20% situs inversus patsientidest



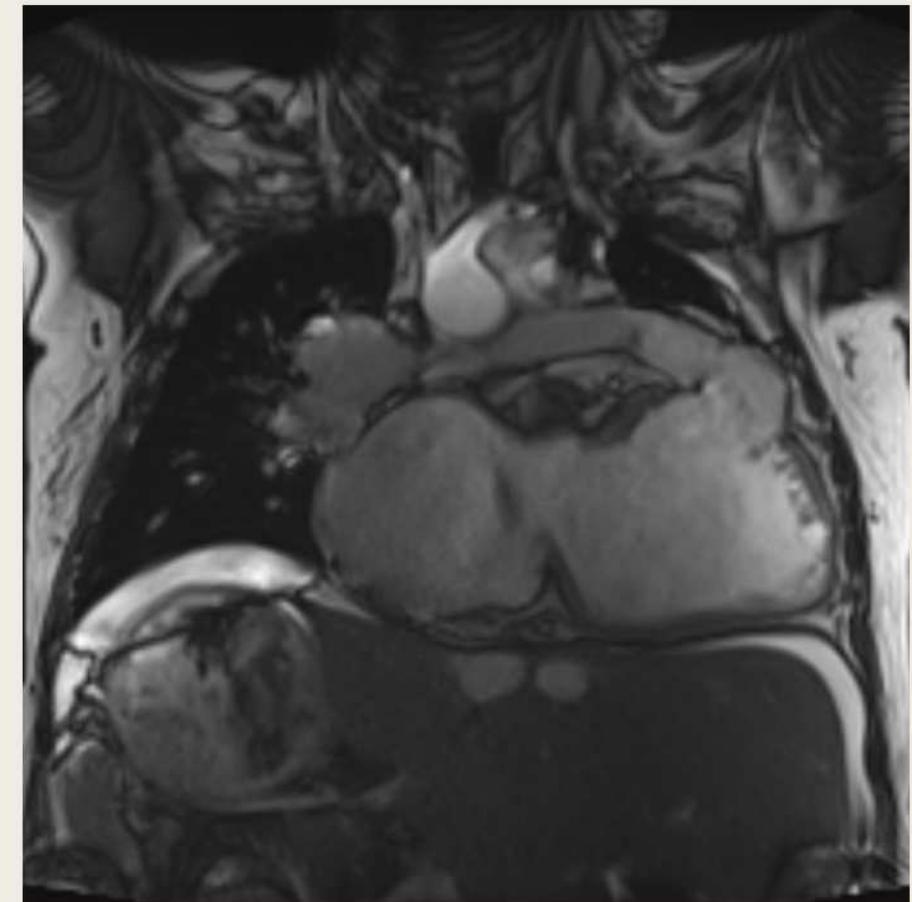
32 a. mees

Situs inversus levocardia ga

1: 2 000 000



PERH 05/17



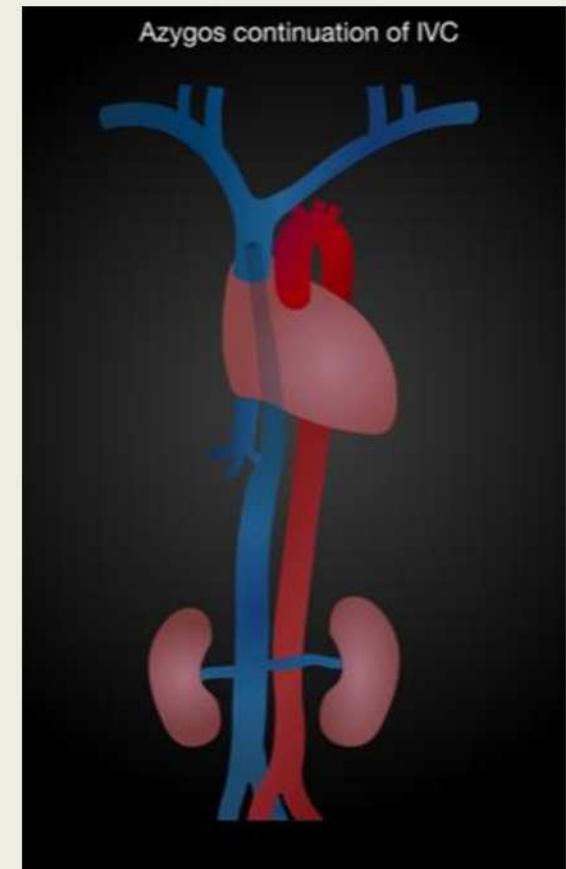
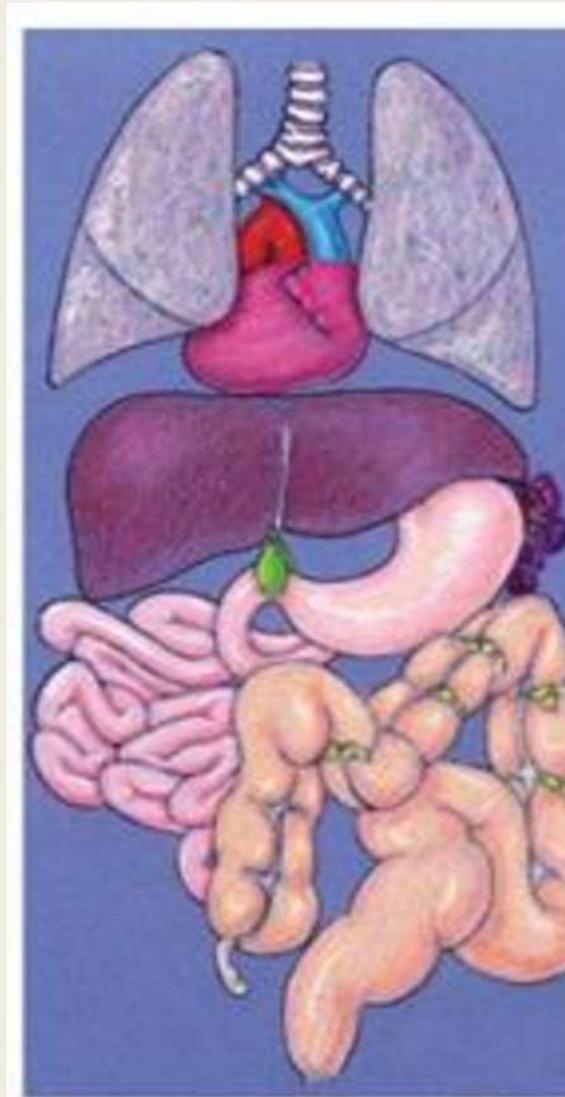
PERH 11/17

Situs ambiguous

- Heterotaxy – abnormaalne organite positsioon, solituse ja inversuse vahepealne variant, organite duplikatsioon.
- Esinemissagedus ~1:20 000
- Ei ole spetsiifilist leidu, asendi anomalaiate erinevad variatsioonid
 - *polysplenia*
 - *asplenia*

Polysplenia/left isomerism/bilateral left-sidedness

- Mitu põrna, keskjoone maks, mõlemas kopsus 2 sagarat, alumise õõnesveeni katkenemine.
- F>M
- Vähem südamerikkeid (50-90%) ja immunodefitsiiti, parem prognoos
- Rohkem anatoomilisi/kliinilisi variatsioone



Interrupted IVC with Az/HemiAz continuation

Asplenia/Right isomerism/bilateral right-sidedness/Ivemark syndrome

- Asplenia (rudimentaарne põrn), keskjoone maks, ipsilateraalne aort ja IVC, mõlemas kopsus 3 sagarat.
- M>F
- 99%–100% kaasasündinud südamerikad, tsüanoos, immunodefisiit
- Kõrge suremus, 1. eluaasta jooksul 80-95%
- Haruldane täiskasvanutel.

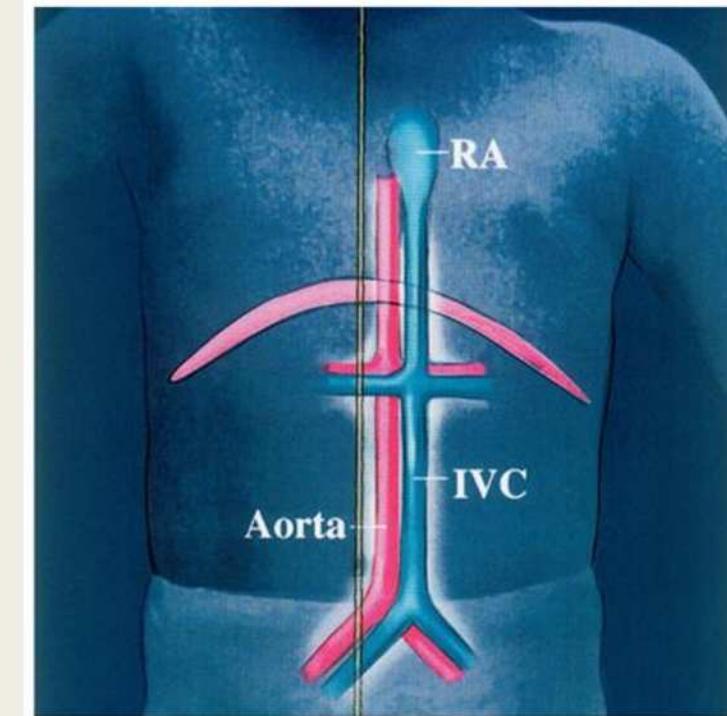
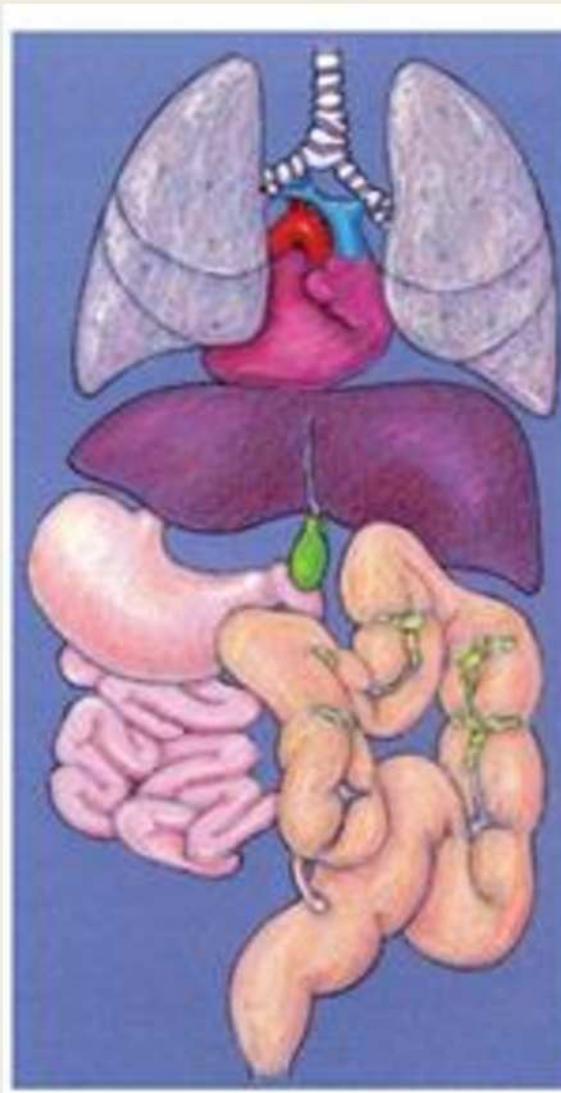
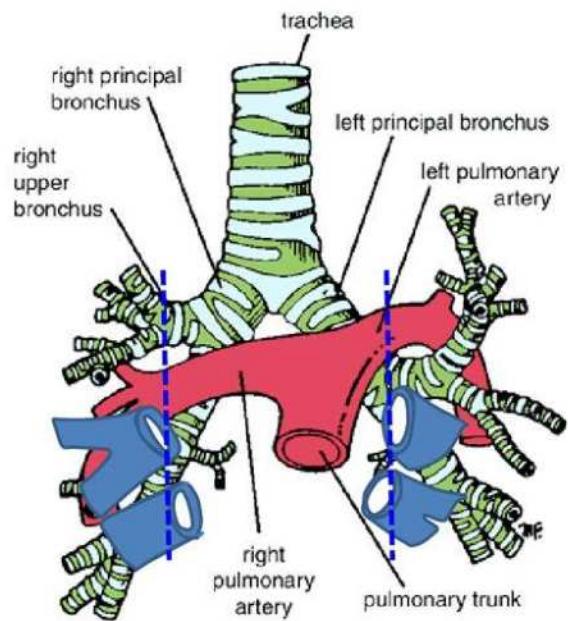


Figure 14. Drawing illustrates the ipsilateral position of the abdominal aorta and inferior vena cava (IVC) classically described as a specific sign of asplenia.

Kimberly E Applegate "Situs revisited: imaging of the heterotaxy" Radiographics 1999

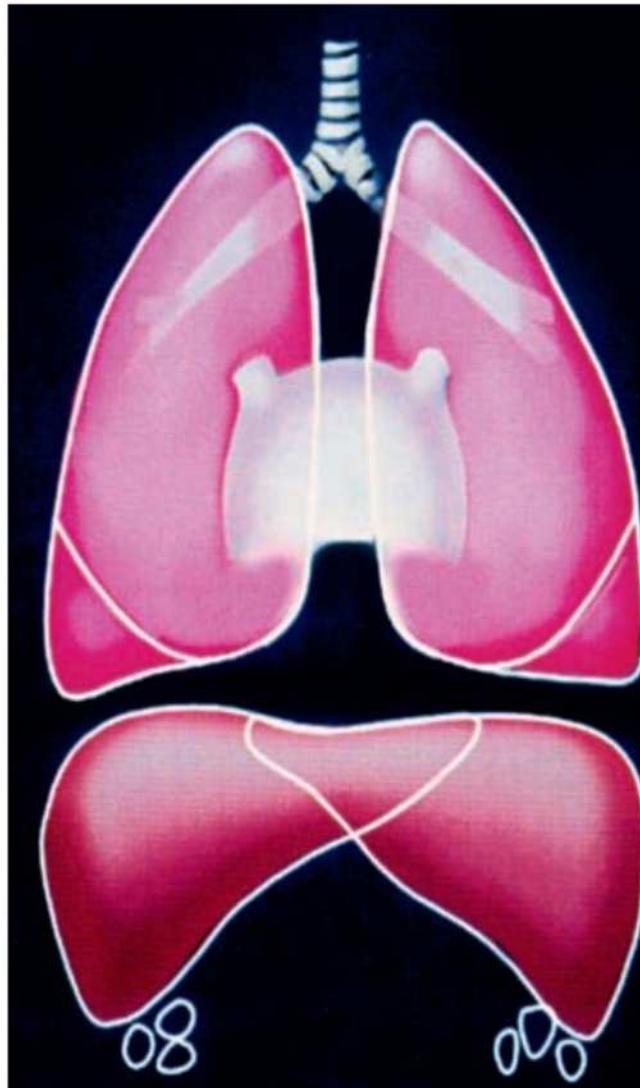
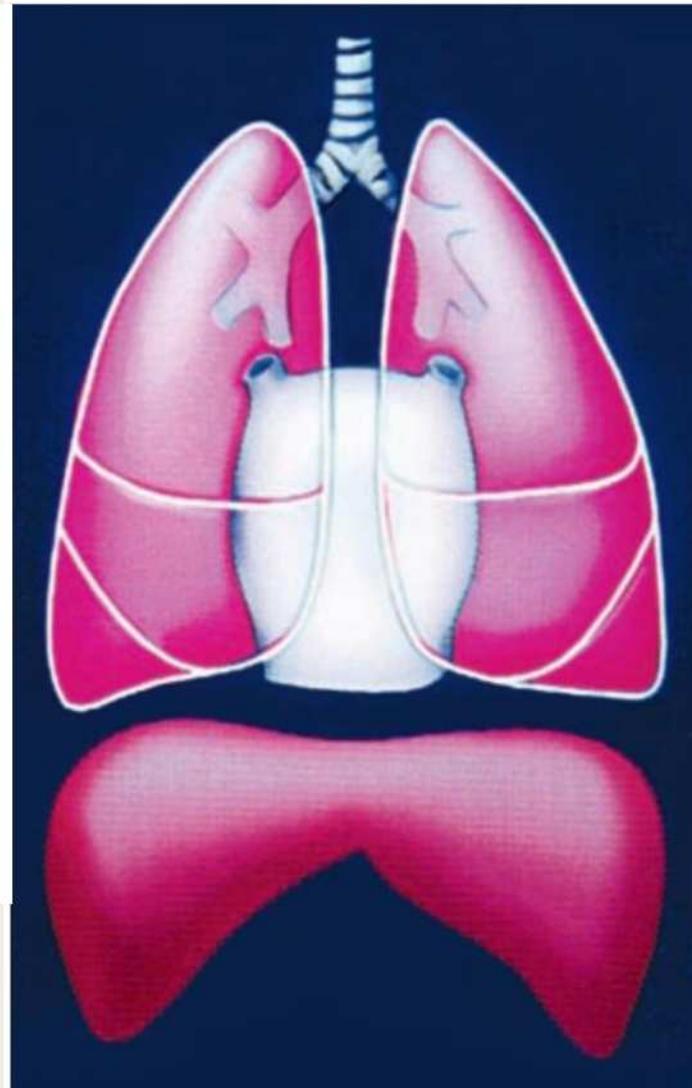
Eparterial bronchi (Asplenia)



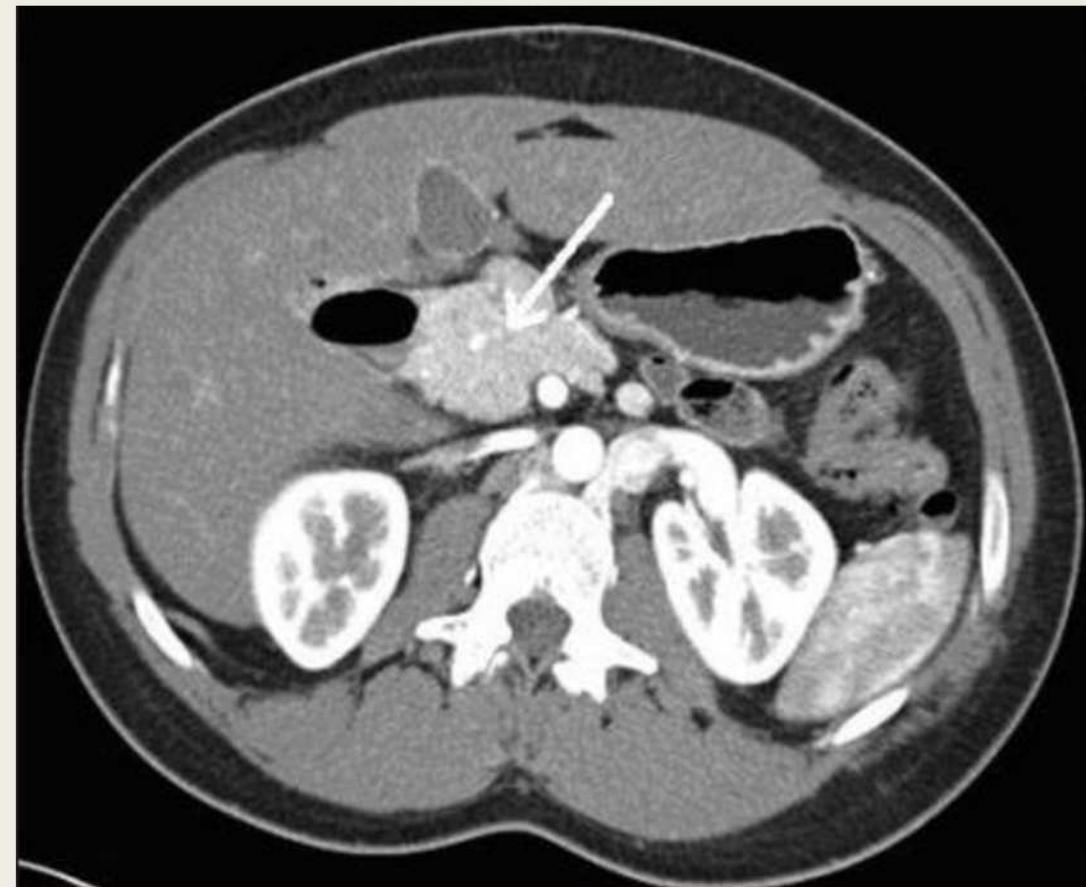
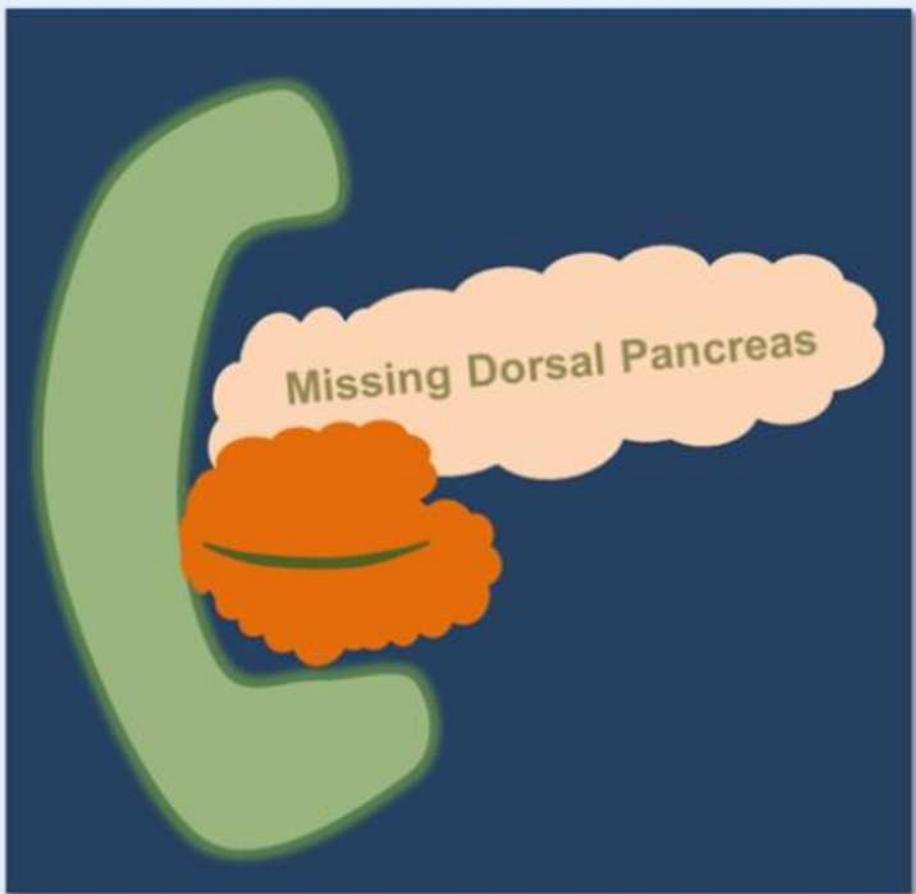
The eparterial bronchus is the right superior lobar (secondary) bronchus; all of the other bronchi are hyparterial bronchi.

<http://slideplayer.com/slide/7221243/>

Hyparterial bronchi (Polysplenia)



Truncated pancreas – pankrease dorsaalne agenees



A.Arora "God Loves Variety Even if We Don't" ECR 2013

Anatomic Characteristics of 21 Patients with Heterotaxy Syndrome

Characteristic, Structure	Patients with Asplenia											Patients with Polysplenia									
	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21
Sex	F	F	M	M	M	M	F	F	M	F	F	F	F	F	F	F	F	F	F	M	
Aortic arch	R	R	R	L	L	L	L	L	L	L	R	R	L	L	R	L	R	L	R	L	
Cardiac apex	R	R	R	L	L	L	M	L	L	L	R	L	L	L	L	R	L	L	L	L	
Stomach	R	L	R	R	R	L	R	L	R	R	L	R	L	R/M	R	R	L	R	R	L/M	
IVC	L	L	L	L	L	R	L	L	L	R	R	L	L	...	R	L	R	L	
Azygous/ hemiazygous continuation	No	No	No	No	No	No	No	...	No	No	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No	Yes
Intestinal malrotation	Yes	Yes	...	Yes	Yes	...	Yes	...	Yes	...	Yes	Yes
Liver	R/M	L/M	M	L/M	M	L/M	M	M	M	R/M	R	M	L/M	R	M	R	R	R	L	L/M	L/M
Gallbladder	R	...	R	M	R/M	...	L/M	L	...	M	...	R	R	R	L	M
Spleen*	0	0	0	0	0	0	0	0	0	0	L(1)	L(3)	R(1)	L(3)	R(1)	R(1)	L(1)	L(1)	R(4)	R(2)	L(1)
Trilobed lungs†	...	Yes	...	Yes	...	R	Yes	R	R
Bilobed lungs†	Yes	Yes	...	Yes	L
Congenital heart disease‡	TA, PS	DORV, AVC	SV, AVC	SV, AVC	TGA, AVC	DORV, AVC	DORV, AVC	Yes	TGA, PS	PA	DORV, HLHS	AVC	AVC	No	No	CA	SV, TGA	LTGA, PA	AVC	No	
Other	BIL SVC	BIL SVC	Fontan	...	L SVC, Fontan	BIL	Fontan	...	L	...	TR	L SVC	BIL SVC

Note.—When the information is unavailable or unknown, the entry is left blank. ASD = atrial septal defect, AVC = atrioventricular canal, BIL = bilateral, CA = common atrium, DORV = double outlet right ventricle, HLHS = hypoplastic left heart syndrome, IVC = inferior vena cava, L = left, LTGA = corrected transposition, M = midline, PA = pulmonary atresia, PS = pulmonic stenosis, R = right, SV = single ventricle, SVC = superior vena cava, TA = tricuspid atresia, TGA = transposition of great arteries, TR = transplant.

*Number in parentheses indicates the number of spleens present.

†As determined with chest radiographic analysis.

‡This list is not inclusive but highlights the major abnormalities in each patient.

From the Department of Radiology, Section of Pediatric radiology,
Cleveland Clinic Childrens Hospital, Ohio.
Radiographics 1999

Imaging Findings in 19 Adults with Situs Anomalies

Anomaly	Situs Inversus (n = 10)										Situs Ambiguous with Polysplenia (n = 8)								Situs Ambiguous with Asplenia (n = 1)
	1/F CT	2/F CT	3/M CR	4/M UGI, CR	5/M MR, Ang	6/M CT, CR	7/M IC	8/F MR	9/M CR	10/M CT, CR	11/M CT, UGI, CR	12/F CT	13/F CT, CR	14/M CT	15/M UGI, BE, US, CR	16/F CT	17/M CT	18/F MR	
Patient no./sex	1/F CT	2/F CT	3/M CR	4/M UGI, CR	5/M MR, Ang	6/M CT, CR	7/M IC	8/F MR	9/M CR	10/M CT, CR	11/M CT, UGI, CR	12/F CT	13/F CT, CR	14/M CT	15/M UGI, BE, US, CR	16/F CT	17/M CT	18/F MR	19/M CT, Nucl
Examinations analyzed																			
Anatomic location																			
Liver	L	L	L	L	L	L	L	L	L	ML	R	ML	ML	ML	ML	R	R	ML	
Spleen*	R	R	R	R	R	R	R	R	R	L(1)	L(10)	R(4)	R(3)	R(>10)	L(8)	L(7)	L(6)	Abs	
Stomach	R	R	R	R	R	R	R	R	R	R	L	R	R	R	L	L	L	R	R
Gallbladder	L	L	NA	NA	L	L	L	NA	L	ML	R	ML	ML	ML	ML	R	R	ML	
Aorta	R	R	NA	NA	R	R	NA	R	NA	R	L	L	L	L	R	L	L	R	R
IVC	L	L	NA	NA	L	L	NA	L	NA	R	R	R	R	R	Dup	R	R	L	L
Cardiac apex	R	R	R	R	R	R	R	R	R	R	L	L	L	L	R	L	L	R	R
Aortic arch	R	R	R	R	R	R	R	R	R	R	I	L	L	L	R	L	I	R	R
Appearance of pancreas	Nor	Nor	NA	NA	Nor	Nor	Nor	Nor	NA	Nor	Nor	TH	NA	TH	TH	TH†	TH‡	TH	TH
Presence of abdominal abnormality												Y	Y	Y	Y	Y	Y	Y	Y
Azygous or hemizygous continuation	N	N	NA	NA	N	N	NA	N	NA	N	N	Y	Y	Y	Y	Y	Y	Y	Y
Small bowel rotational abnormality	NA	Y	NA	Y	Y	Y	Y	Y	NA	Y	Y	Y	N	Y	Y	Y	Y	Y	Y
Colon reverse rotational abnormality	NA	Y	NA	Y	Y	Y	Y	Y	Y	Y	Y	Y	N	Y	Y	Y	Y	Y	Y

Abbreviations: Abs = absent, Ang = angiography, BE = barium enema study, CR = conventional radiography, Dup = duplicated, IC = intraoperative cholangiography, L = left, ML = midline, N = no, NA = not available, Nor = normal, Nucl = nuclear medicine study, R = right, TH = truncated (pancreatic) head, UGI = upper gastrointestinal series, Y = yes.

Note.—Congenital heart disease was not seen in any patient.

*Numbers in parentheses indicate number of spleens.

†Midline (in all other affected patients, the truncated head was on the right).

‡Clefted.

From the Department of Radiology, Medical College of Virginia of Virginia Commonwealth University, 401 N 12th St, Rm 3-407B, Richmond

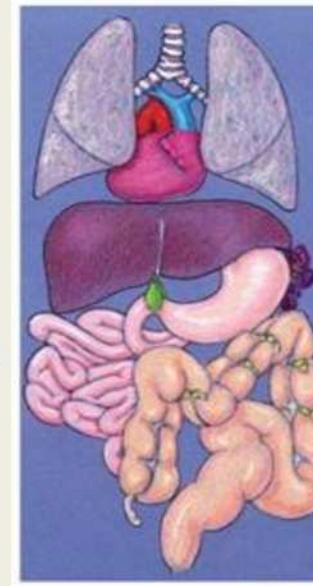
Radiographics 2002

Situs ambiguus

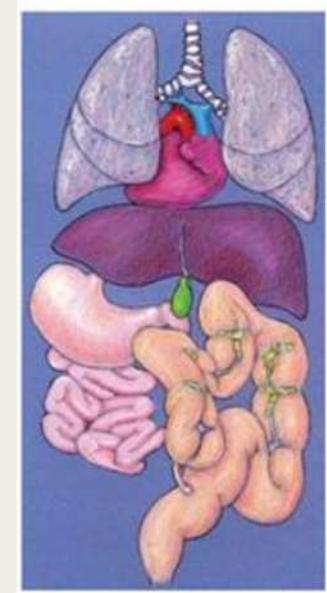
Kokkuvõte:

- Inversuse ja solituse vahepealne variant , varieeruv anatoomia, võib simuleerida teisi haigusi
- Meie patsiendil: Heterotaaksia sündroom parempoolse polüspleeniaga ja maoga, levokardiaga, vasakpoolse ipsilateraalse alumise õõnesveeniga.
- UH – IVC asend!
- Edasised uuringud: Ehho, rindkere KT, südame KT/MRT?

Polysplenia >



Asplenia



Kasutatud kirjandus:

- Ann S. Fulcher, Mary Ann Turnen "Abdominal manifestations of situs anomalies in adults" Radiographics 2002; 22:1439-1456
- Kimberly E Applegate, Marilyn J. Goske, Gregory Pierce, Daniel Murphy "Situs revisited: imaging of the heterotaxy" Radiographics 1999; 19:837-852
- <https://radiopaedia.org/articles/situs-inversus>
- <https://radiopaedia.org/articles/heterotaxy-syndrome>
- <https://radiologykey.com/cardiosplenic-syndromes/>
- A.Arora "God Loves Variety Even if We Don't" ECR 2013
http://posterng.netkey.at/esr/viewing/index.php?module=viewing_poster&task=viewsection&pi=114841&ti=370421&searchkey=
- <https://radiopaedia.org/articles/kartagener-syndrome-1>