# Neerude tsüstiliste haiguste käsitlus

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# Sissejuhatus

- Üle 50 aastaste seas UH juhuleiuna vähemalt 50%-l neerudes tsüst(id)
- Klassifitseerimine:
  - Põhjus: geneetiline või omandatud
  - Tsüsti välimus: lihtne või komplitseeritud
  - Asukoht: kortikaalne või medullaarne

WeberTM. Ultrasound Clin 2006 : 1 : 15-24.

# Põhilised haigused

- Omandatud:
  - Lihtne tsüst; omandatud tsüstiline neeruhaigus; *medullary sponge kidney*
- Geneetilised:
  - ARPKD (*autosomal recessive polycystic kidney disease*); ADPKD (*autosomal dominant polycystic kidney disease*); JNPHP (*juvenile nephronophthisis*); MCKD (*medullary cystic kidney disease*)
- Süsteemsete haigustega seotud neerutsüstid:
  - Von Hippel-Lindau sündroom; tuberoosne skleroos
- Maliigsus:
  - Tsüstiline neerurakuline vähk

# Epidemioloogia

- ADPKD 1:400-1000, meestel varasema algusega
- ARPKD 1:6000-55000, heterosügootse kandja sagedus 1:70
- Tuberoosne skleroos 1:10000-50000
- Von Hippel-Lindau sündroom 1:39000 – 2/3 neerude tüstid 30-40 aastaselt
- *Medullary sponge kidney* 1:5000 (M:N 2:1), 20% lisandub nefrolitiaas (kaltsiumkivid)

# Bosniak klassifikatsioon

- Hindab tsüstilisi neerude masse
- Töötati välja KT-uuringute põhjal 1986.a. ning täiendati 2005.a.
  - Osa II kategooria tüstide maliiguses oli kuni 20%
- Mõeldud radioloogiliseks ja kliiniliseks hindamiseks
  - Hiljem kanti üle ultraheli ja MRT uuringutele
- Maliigsus
  - I- 0%
  - II- 0%
  - IIF- kuni 20%
  - III- 50%
  - IV- 100%
-

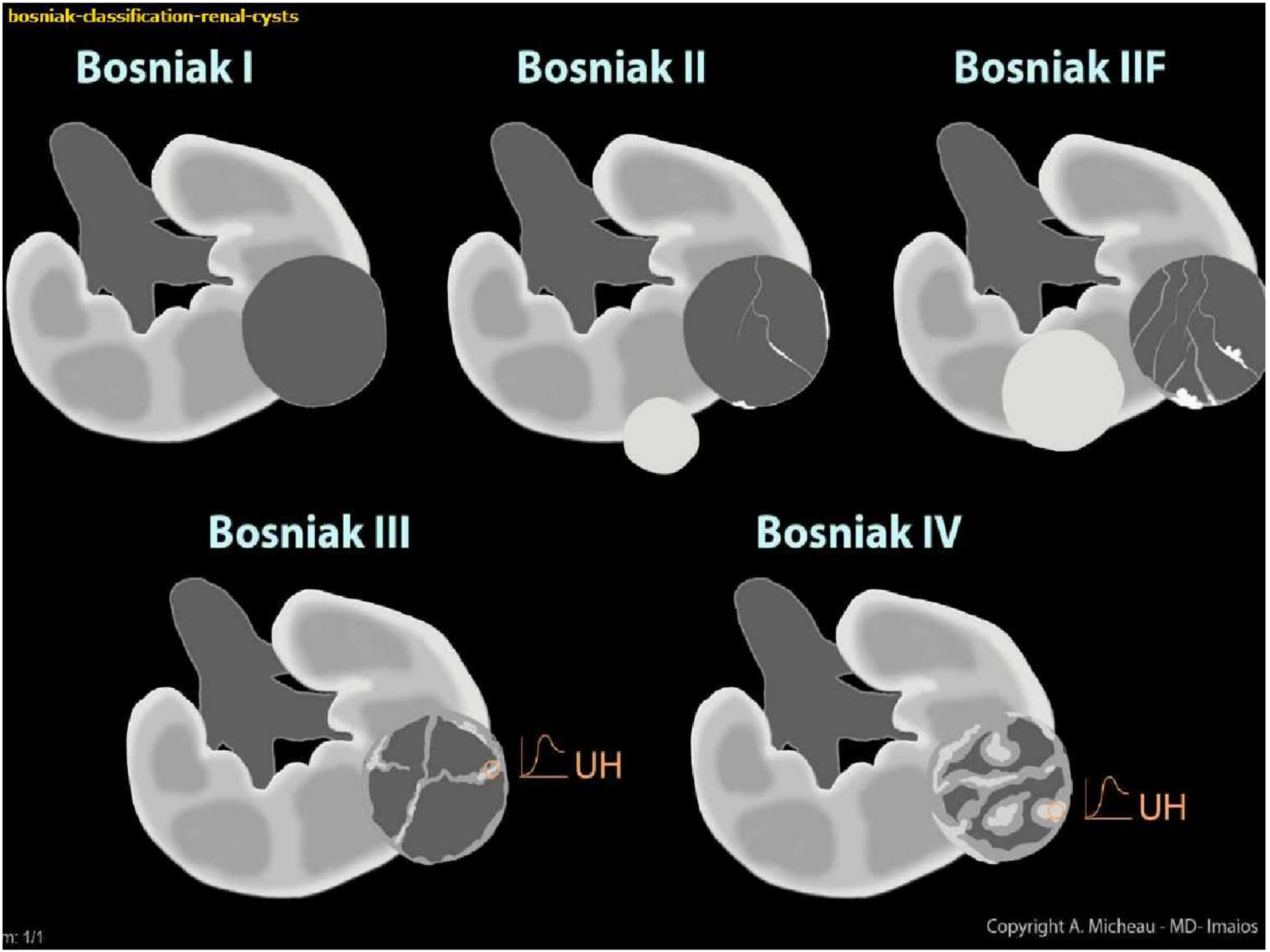
| Bosniak category | Features |
|---|---|
| I | A simple benign cyst with a hairline thin wall that does not contain septa, calcification or solid components. It measures as water density and does not enhance with contrast material. |
| II | A benign cyst that might contain a few hairline thin septa. Fine calcification might be present in the wall or septa. Uniformly high-attenuation lesions of <3 cm that are sharply marginated and do not enhance. |
| IIF | These cysts might contain more hairline thin septa. Minimal enhancement of a hairline thin septum or wall can be seen and there might be minimal thickening of the septa or wall. The cyst might contain calcification that might be nodular and thick but there is no contrast enhancement. There are no enhancing soft-tissue elements. Totally intrarenal non-enhancing high-attenuation renal lesions of ≥3 cm are also included in this category. These lesions are generally well marginated. |
| III | These lesions are indeterminate cystic masses that have thickened irregular walls or septa in which enhancement can be seen. |
| IV | These lesions are clearly malignant cystic lesions that contain enhancing soft-tissue components. |

# Käsitlus

- I- ignoreeri
- II- ignoreeri
- IIF- jälgi
- III- lõika
- IV- lõika

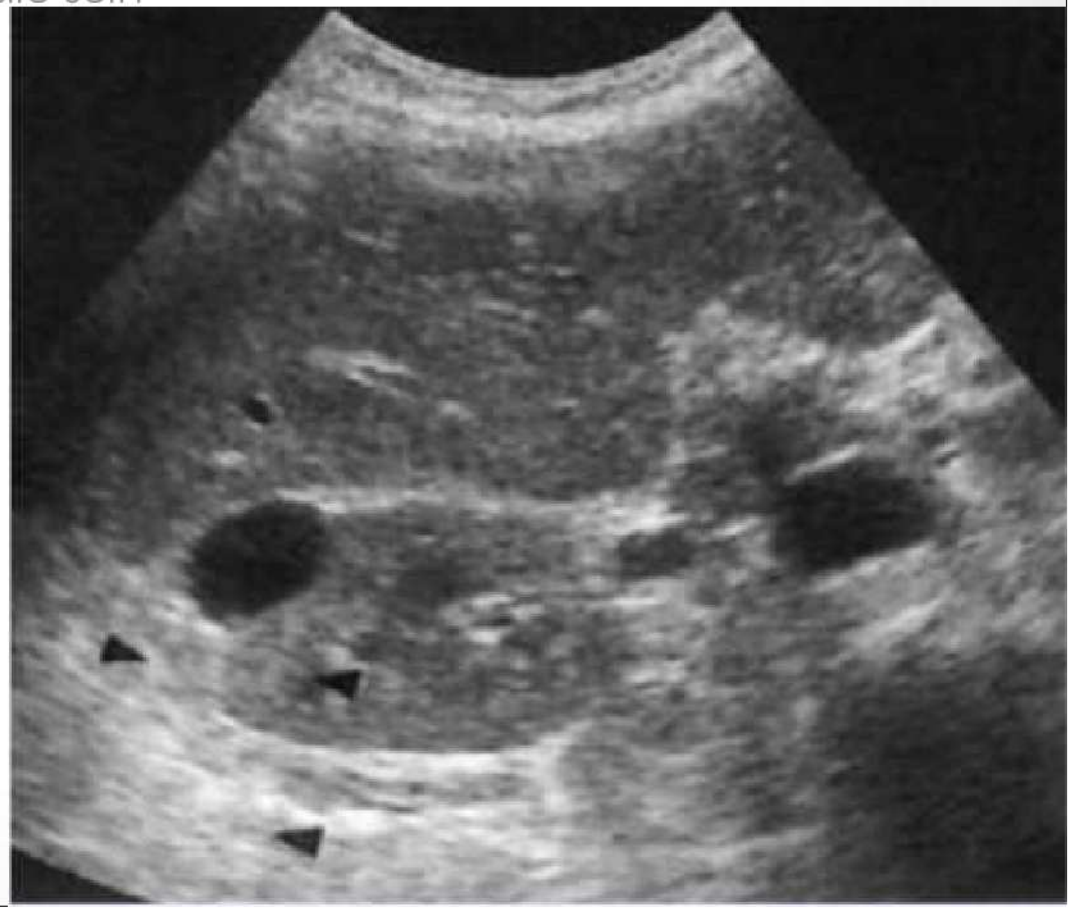
**The Bosniak Renal Cyst Classification System**

| Category | Criteria and Management |
|---|---|
| I | A benign simple cyst with a hairline-thin wall that does not contain septa, calcifications, or solid components; it has water attenuation and does not enhance; no intervention is needed |
| II | A benign cystic lesion that may contain a few hairline-thin septa in which perceived (not measurable) enhancement may be appreciated; fine calcification or a short segment of slightly thickened calcification may be present in the wall or septa; uniformly high-attenuating lesions (<3 cm) that are sharply marginated and do not enhance are included in this group; no intervention is needed* |
| IIF† | Cysts may contain multiple hairline-thin septa; perceived (not measurable) enhancement of a hairline-thin smooth septum or wall can be identified; there may be minimal thickening of wall or septa, which may contain calcification that may be thick and nodular, but no measurable contrast enhancement is present (45); there are no enhancing soft-tissue components; totally intrarenal nonenhancing high-attenuating renal lesions (>3 cm) are also included in this category; these lesions are generally well marginated; they are thought to be benign but need follow-up to prove their benignity by showing stability (46)* |
| III | Cystic masses with thickened irregular or smooth walls or septa and in which measurable enhancement is present; these masses need surgical intervention in most cases, as neoplasm cannot be excluded; this category includes complicated hemorrhagic or infected cysts, multilocular cystic nephroma, and cystic neoplasms; these lesions need histologic diagnosis, as even gross observation by the urologist at surgery or the pathologist at gross pathologic evaluation is frequently indeterminate |
| IV | Clearly malignant cystic masses that can have all of the criteria of category III but also contain distinct enhancing soft-tissue components independent of the wall or septa; these masses are clearly malignant and need to be removed |


bosniak-classification-renal-cysts
Bosniak I
Bosniak II
Bosniak IIF
Bosniak III
UH
Bosniak IV
UH
Copyright A. Micheau - MD- Imaios
m: 1/1

# Bosniak I

- UH:
  - Anehhogeenne
  - Teravalt piirdunud õhuke ja sile sein
  - Kajavõimendus tsüsti taga
- KT:
  - Õhuke sein, kus pole septe, kaltsifikaate ega pehmekoelist komponenenti
  - Vee tihedus
  - Ei kontrasteeru
  - Maliigsus: 0%



# Komplitseeritud neerutsüst

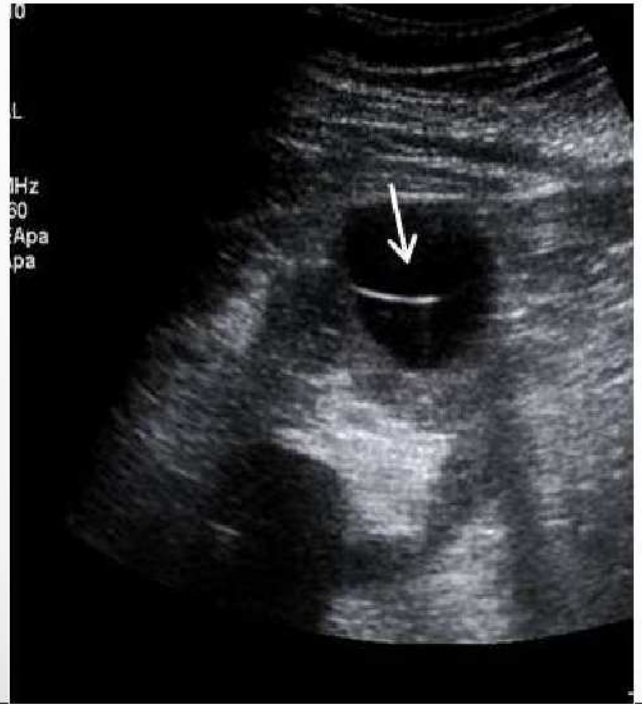
- Seina paksenemine
- Seintest lähtuvad septid ja/või noodulid
- Kaltsifikaadid
- Kõrge tihedus (>20HU KT-uuringul)(vee tihedus kuni 20HU)
- Kontrasteerumine (>20HU erinevust natiiv ja kontrast -uuringul)

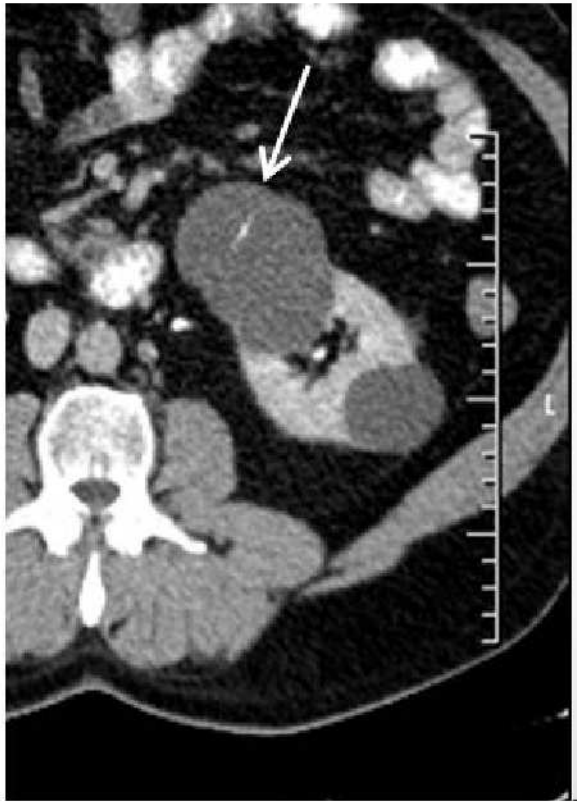
Hartman DS et al. Radiographics 2004 ; 24: S101 – S115

# Bosniak II

- Üksikud õhukesed septid, kus aimatav*(perceived)* kontrastaine kogunemine (pole mõõdetav*)*
- Vähesed õhukesed seina või septi pinna kaltsifikaadid. Veidi paksem kaltsifikaat ainult lühikesel seina segmendil.
- Kõrge tihedusega (>20HU) hästi piirdunud kolded

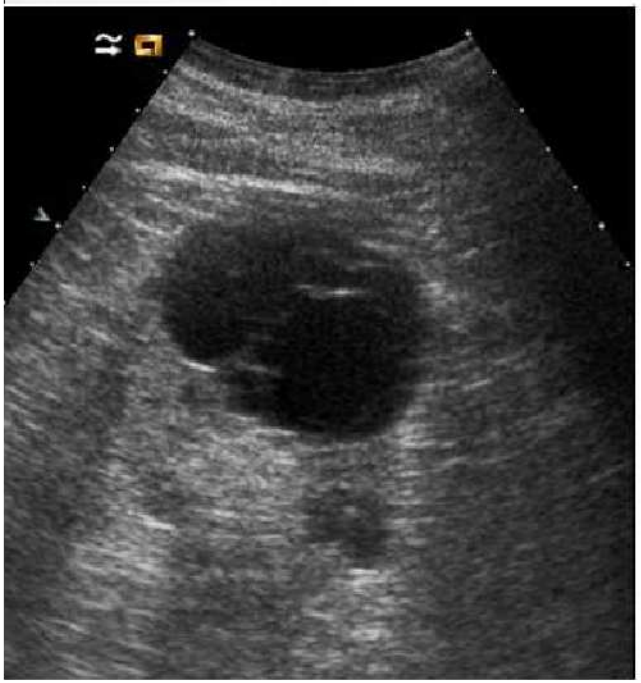
  D<3cm, mis ei kontrasteeru.

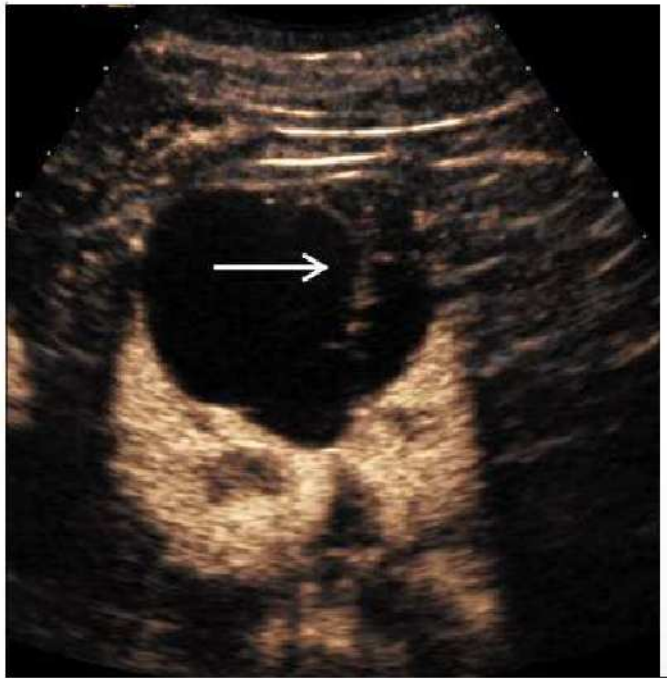




# Bosniak IIF

- Mitmed õhukesed septid, kus võib olla aimatav kontrastaine kogunemine (pole mõõdetav)
- Vähene seinte või septide paksenemine ja paksud või nodulaarsed kaltsifikaadid, kuid puudub mõõdetav kontrasteerumine
- Hästipiirdunud intrarenaalsed kolded tihedusega >20HU ning D>3cm, mis ei kontrasteeru. Kui follow-up jälgimisel ei muutu, võib lugeda healoomulisteks.
- Vajavad jälgimist (soovituslikult kontrast KT)





Vasakul tavaline UH

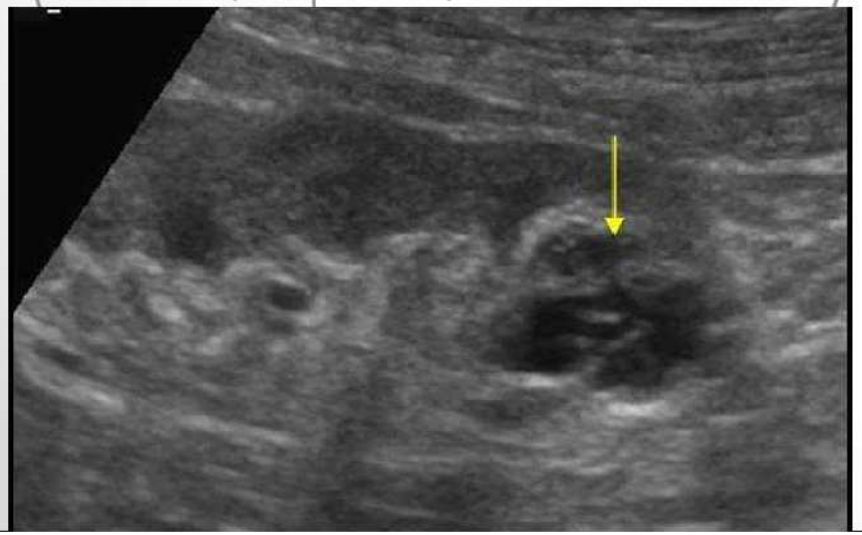
Parempooleslt kontrastainega UH

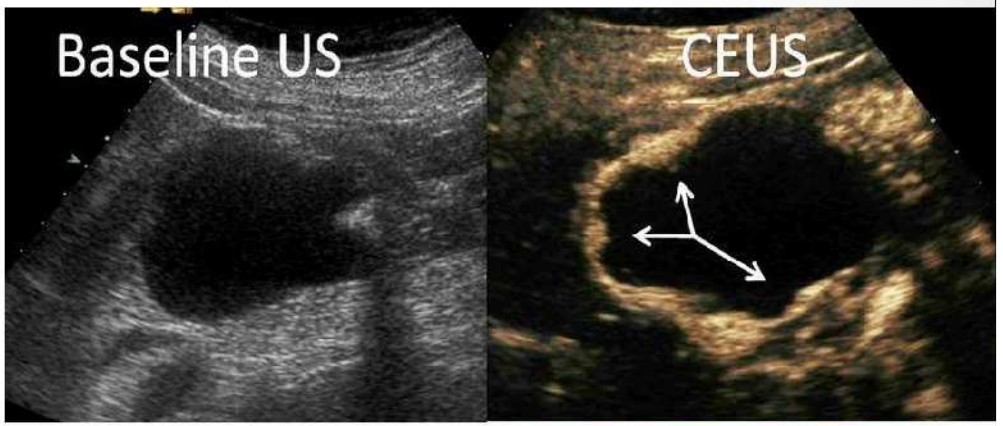
# Bosniak III

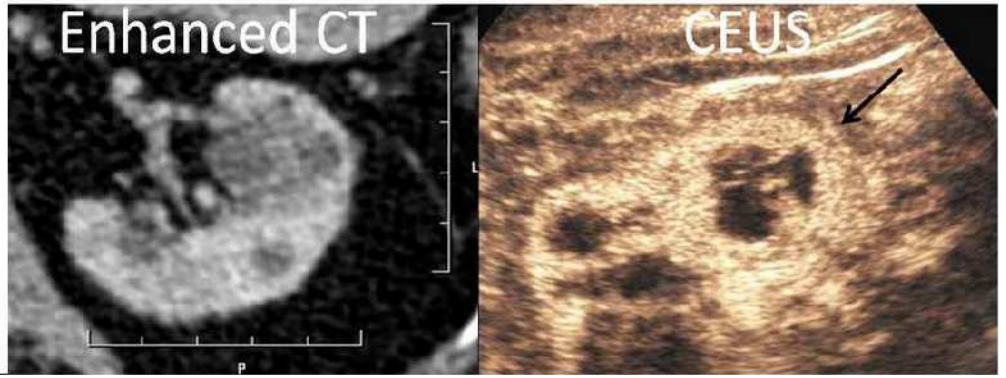
- Paksenenud ebaregulaarsed seinad või septid, kus näha mõõdetavat kontrasteerumist
- Siia kuuluvad ka komlpitseeritud hemorraagilised või infitseeritud tsüstid
- Maliigsus ~50%: vajavad kirurgilist sekkumist
- Biopsia: positiivne vastus kinnitab ->op

negatiivne vastus ei välista maliigsust ->op

Biopsial külvi ja tüsistuste oht (veritsus, ruptuur, infitseerumine)

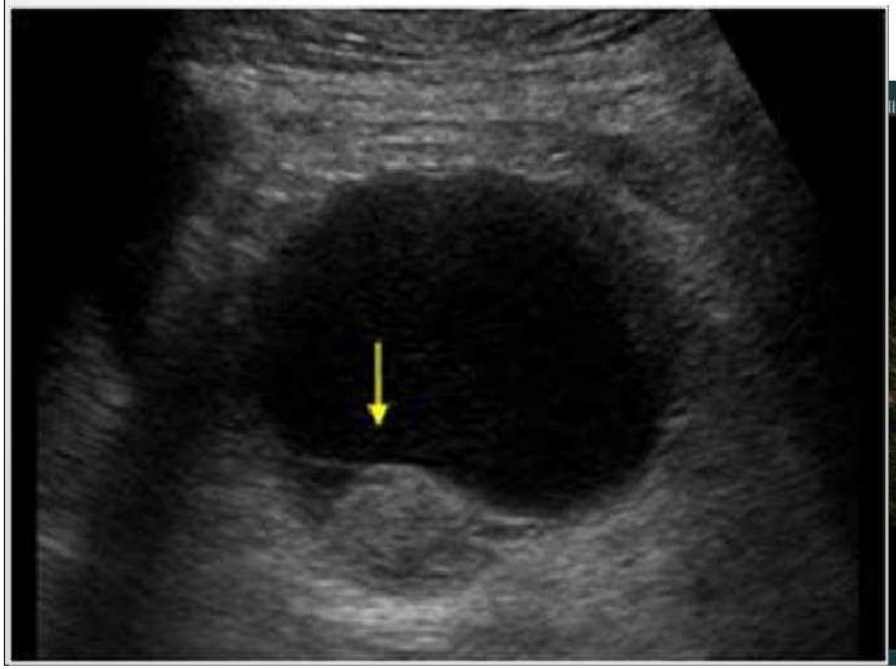


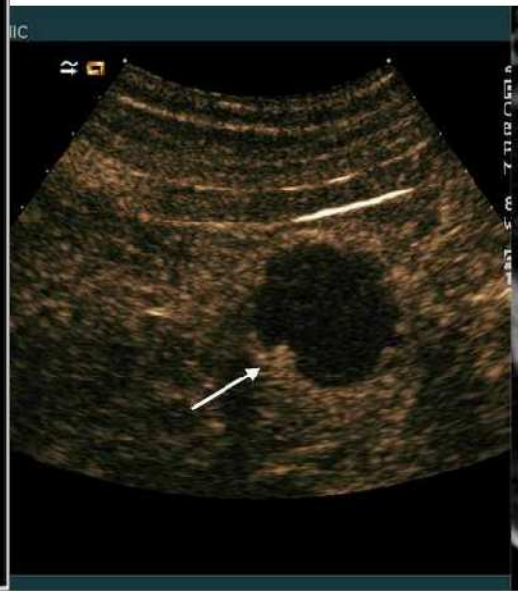


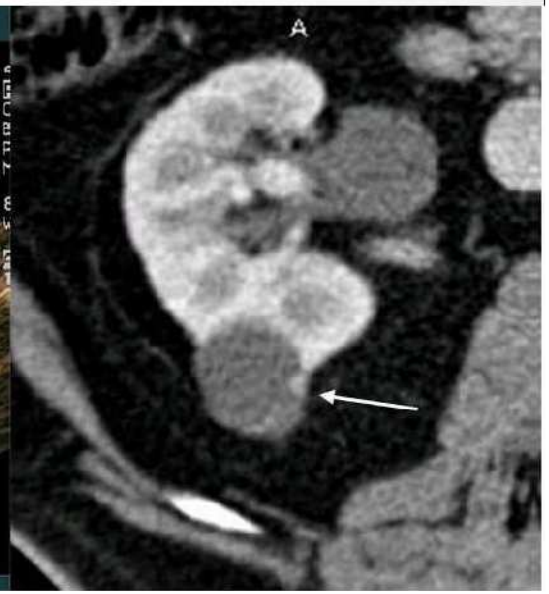



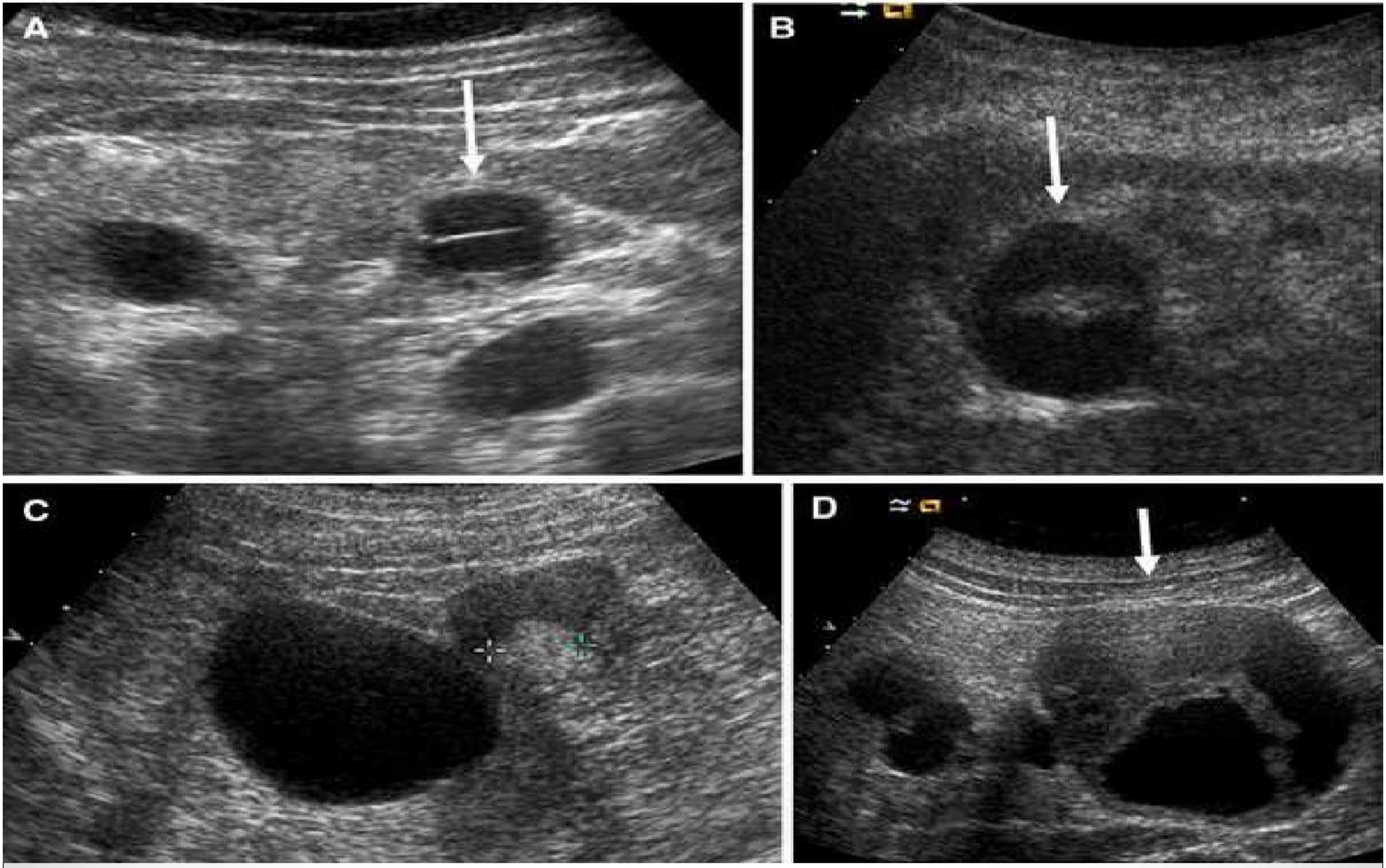
# Bosniak IV

- Paksenenud ebaregulaarsed seinad või septid, kus näha mõõdetavat kontrasteerumist
- Lisaks selgelt eristatav kontrasteeruv pehmekoeline komponent, mis lähtub seinast või septist
- Maliigsus ~100%
- Käsitlus: operatiivne ravi

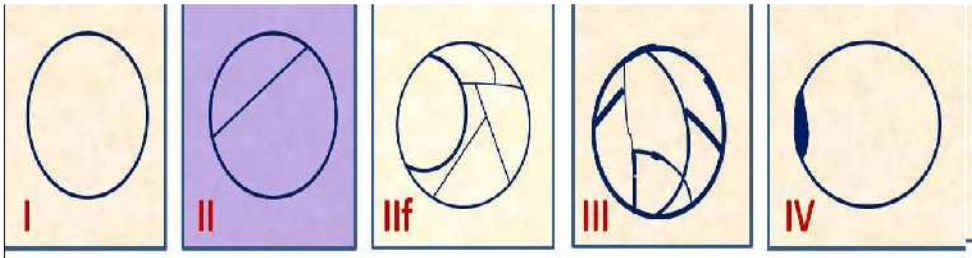


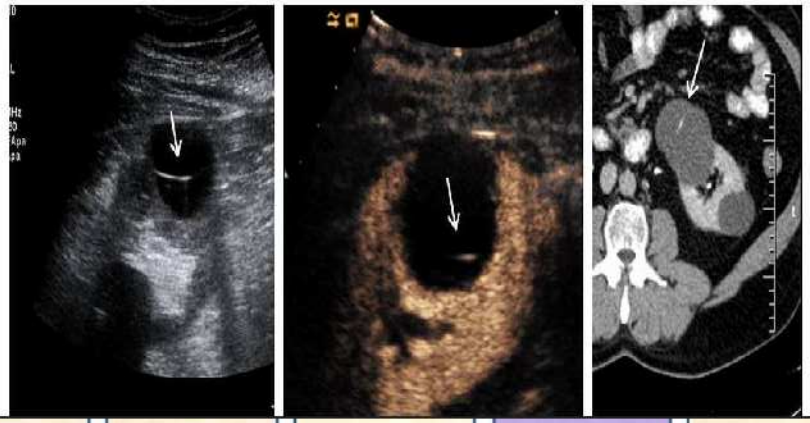


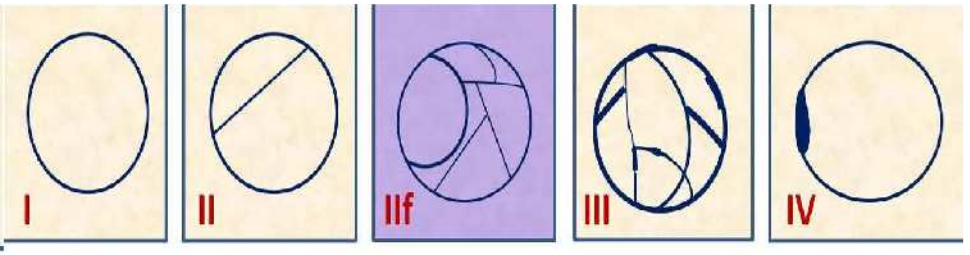





A)Õhuke sept B)Paksenenud sept C)Tsüsti sees noodul D)Hüperehhogeenne sisu tsüsti sees


I
II
IIf
III
IV

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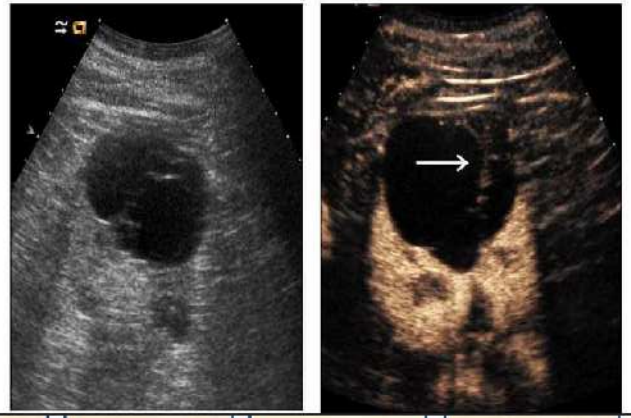
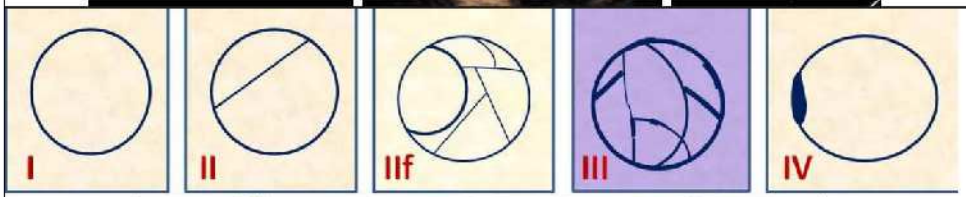
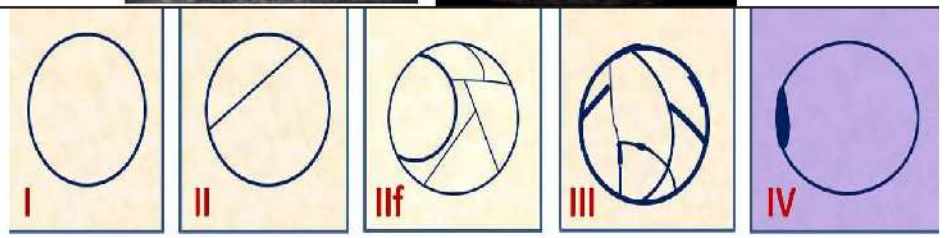
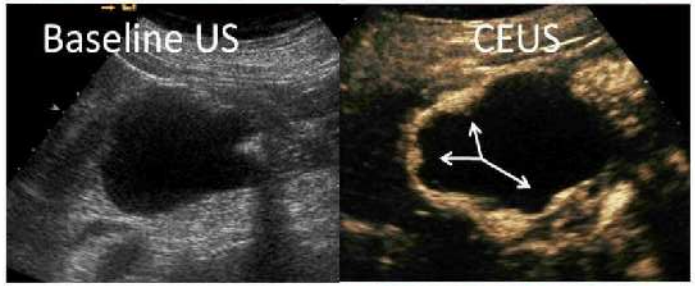
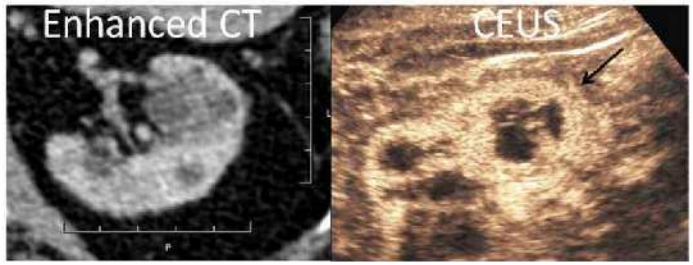
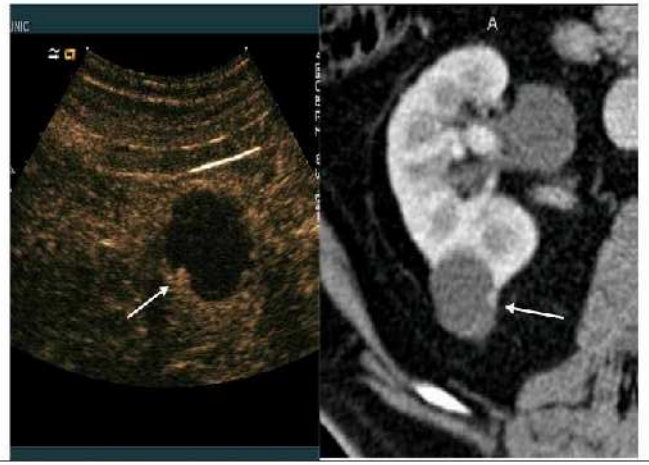



I
II
IIf
III
IV

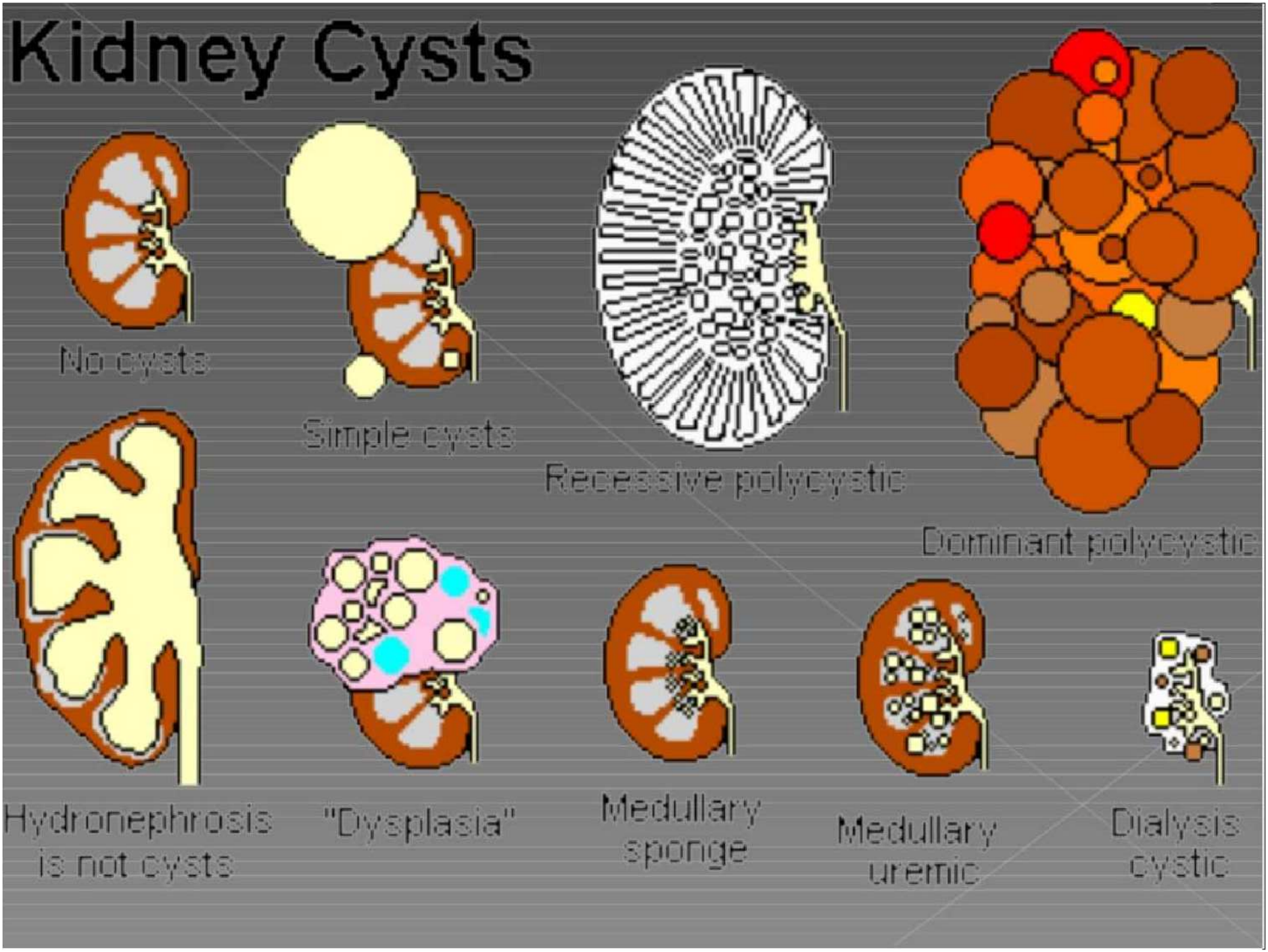

I
II
IIf
III
IV


Baseline US
CEUS


Enhanced CT
CEUS



# Kidney Cysts



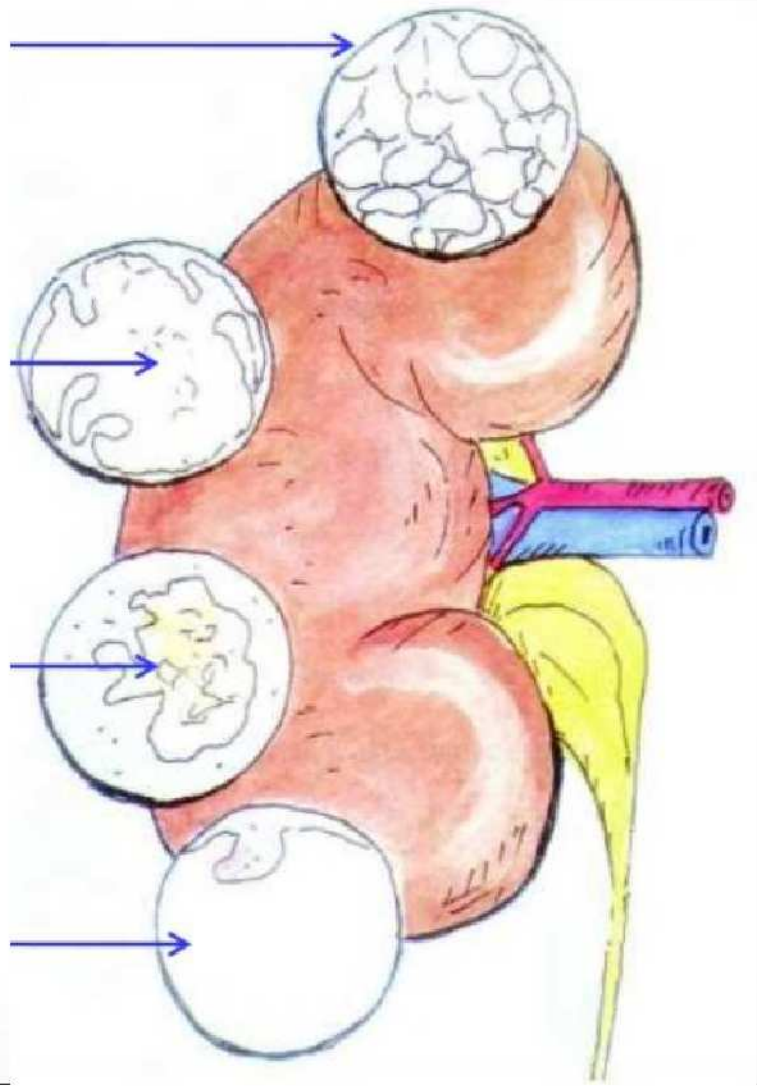
# Neerurakulise vähi tsüstilised kasvumustrid

Multilokulaarne
(3 või enam septi)

Unilokulaarne

Tsüstiline nekroos

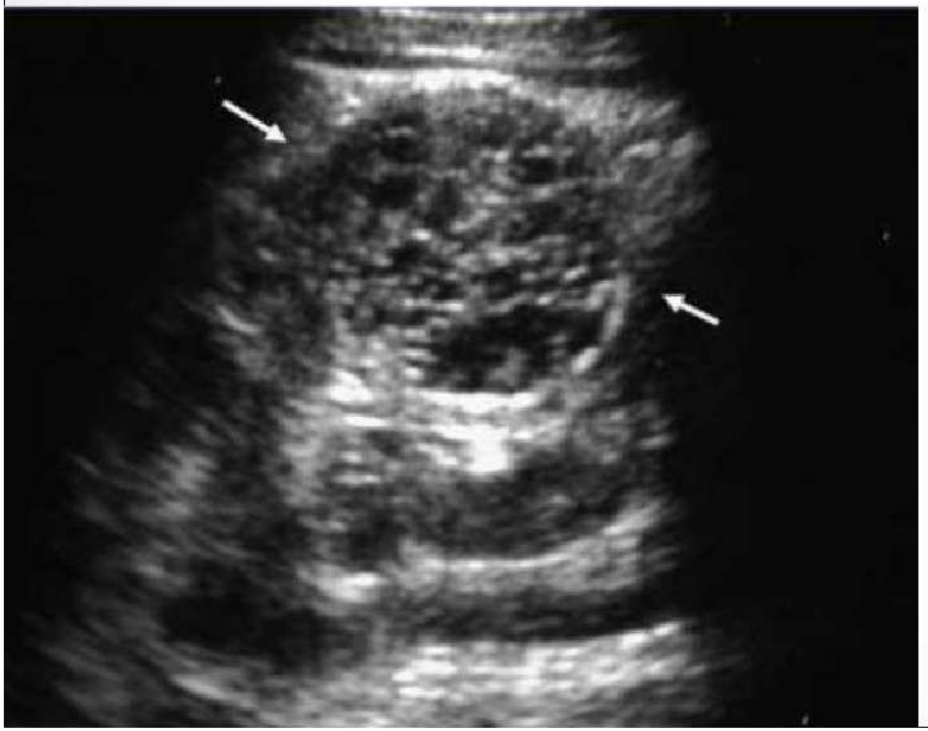
Lihtsa tsüsti seinast lähtuv

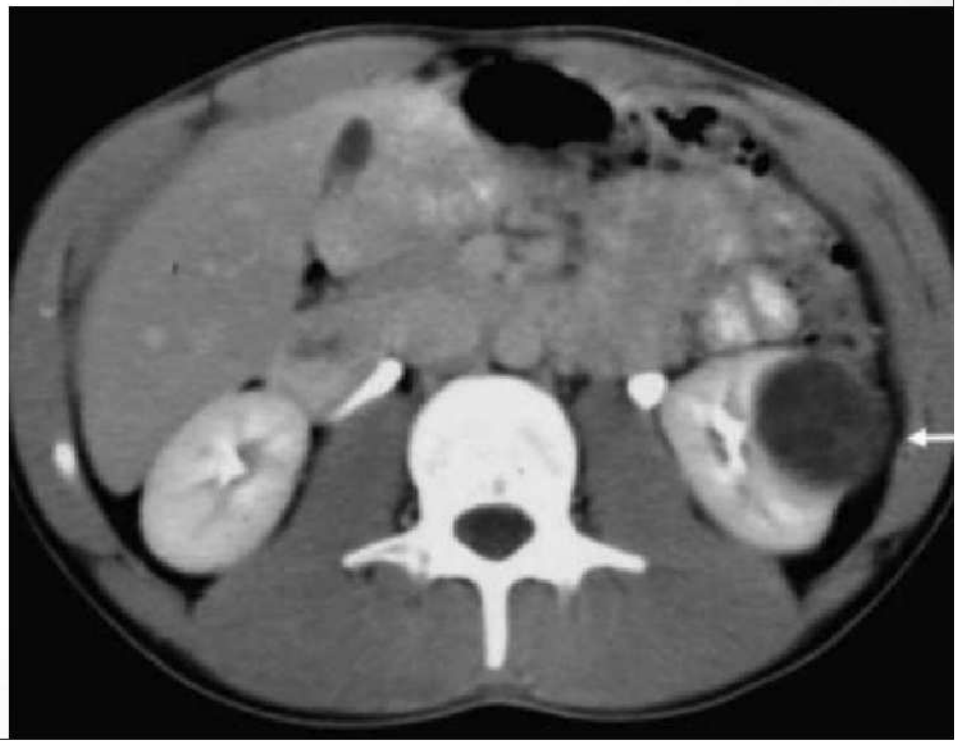


# Multilokulaarne tüstiline nefroom

- Healoomuline pärilik tsüstiline kasvaja
- Mitmed septidega tsüstid, mida pole võimalik eristada multilokulaarsest neerurakulisest vähist ->

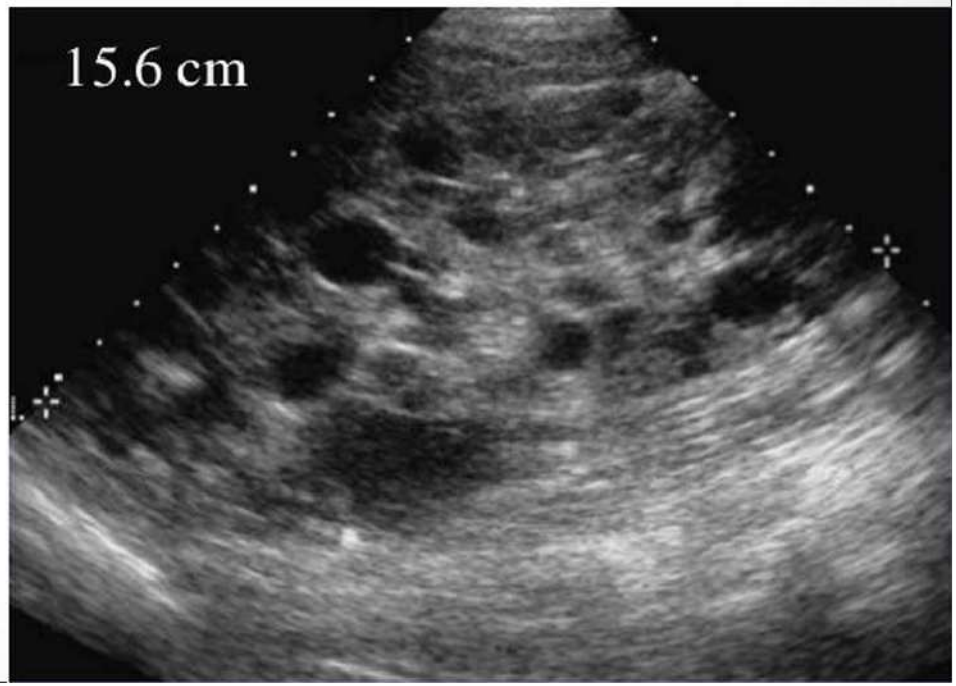
mõlemal juhul operatiivne ravi



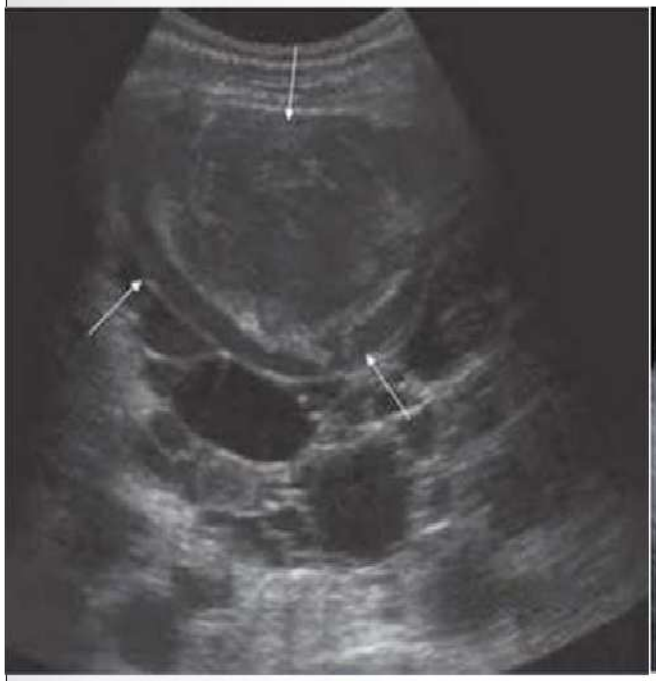


# ADPCKD

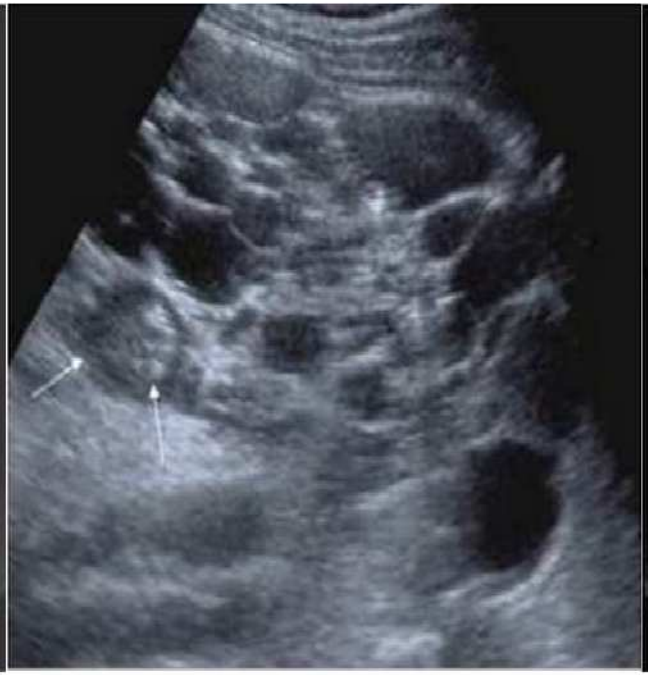
- Bilateraalselt erineva suurusega tsüstid ning suurenenud neerud
- Lisaks maksa, pankrease, arahnoid ja teised tsüstid
- Kriteeriumid:
  - 30. eluaastaks mõlemas neerus vähemalt 2 tsüsti
  - Perekonna anamneesis ADPCKD
  - Tsüstid ka teistes organites
  - Geeni markerid ADPCKD-le



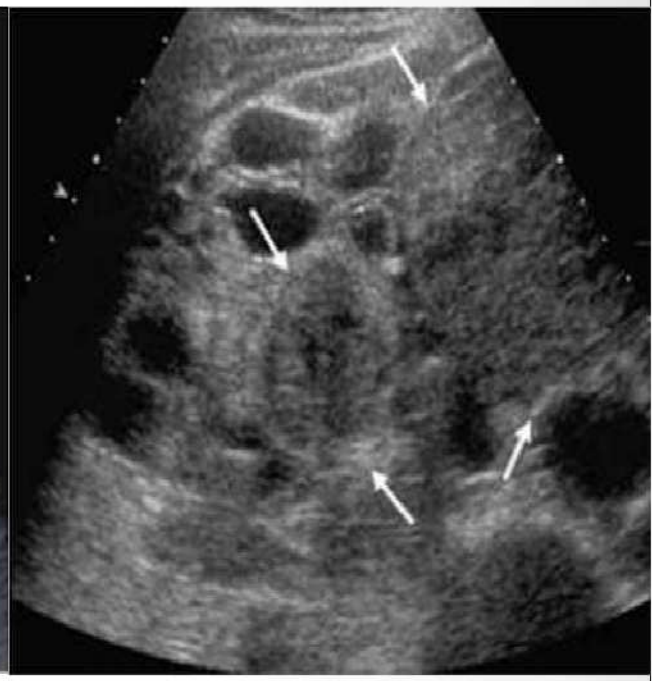
# Komplitseeritud ADPCKD tsüstid



Ehhogeenne tsüst:
Punktsioonil veri



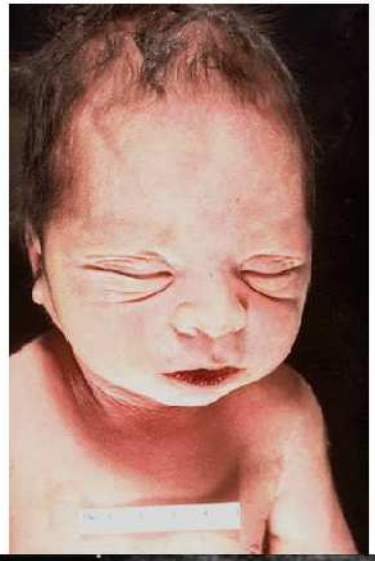
Ehhogeenne tsüst:
Punktsioonil mäda

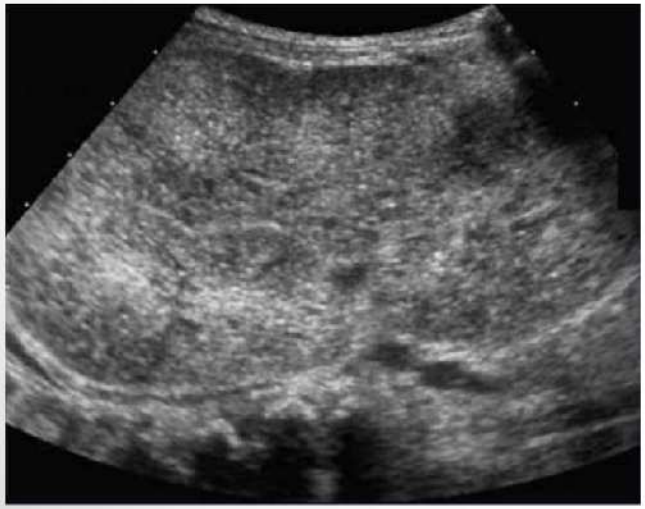


Pehmekoeline mass:
Papillaarne neerurakuline vähk

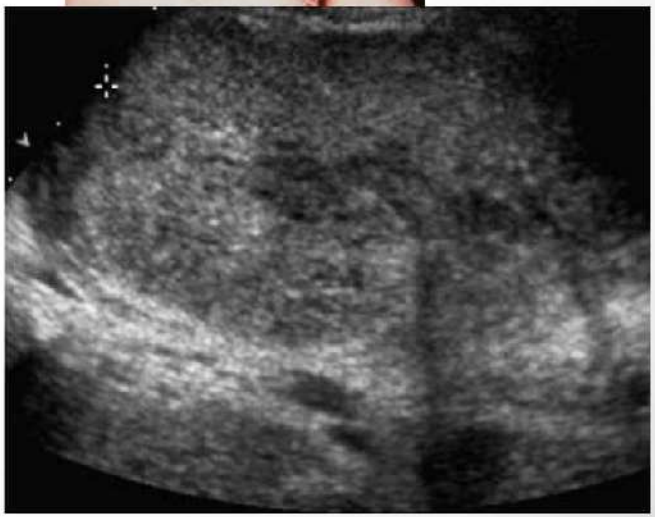
# ARPCKD

- Erineva astme neerupuudulikkus ja hüpertensioon
- Tõsise vormi puhul
  - Peamiselt avaldub neeruhaigus
    - Neerud bilateraalselt suurenenud
    - Potter facies
    - Pulmonaalsed komplikatsioonid
- Hiline avaldumine
  - Neerupuudulikkus
  - Neerud bilateraalselt suurenenud





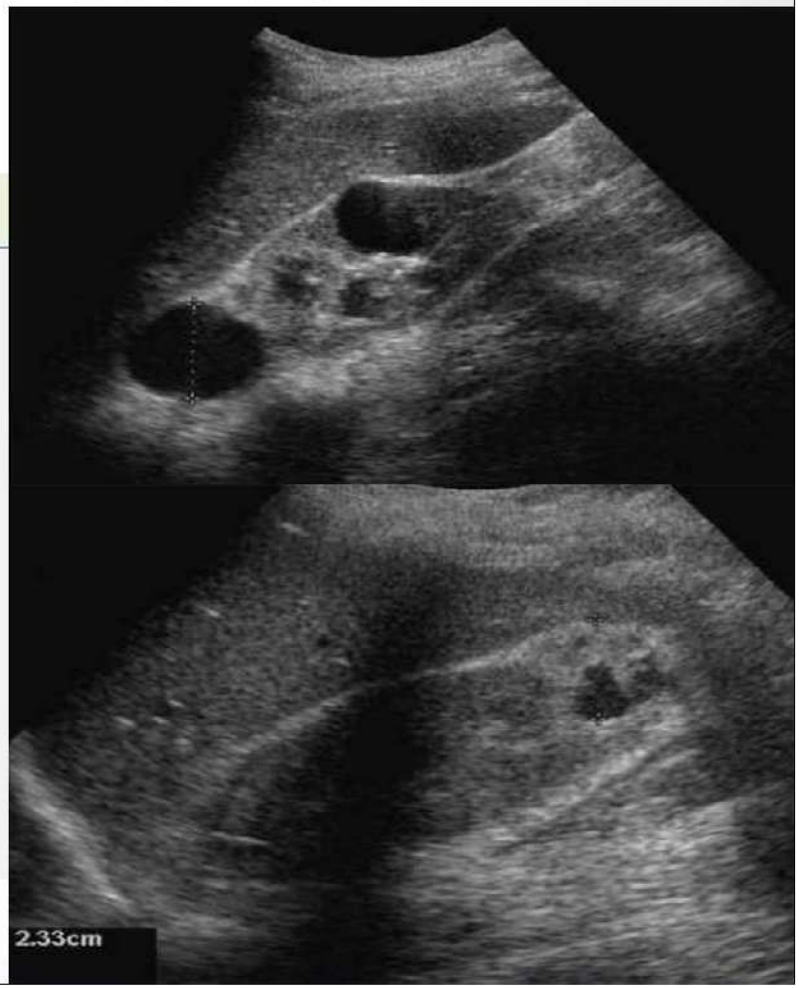
Vastsündinu
<-Parem neer
10cm
Vasak neer ->
9,8cm



# Von Hippel-Lindau (VHL) sündroom

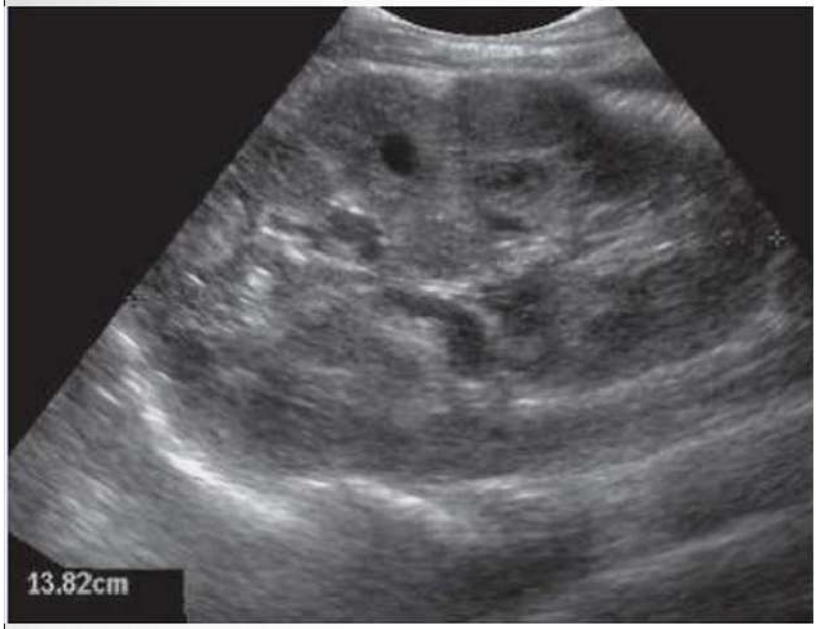
- Autosoom-dominantne haigus
- Eelsoodumus erinevateks hea- ja pahaloomulisteks tuumoriteks
- Ligi 40 kollet 14 erinevas organis

| Manifestations | Prevalence |
|---|---|
| Pancreatic cysts | 50 – 91% |
| Cerebellar hemangioblastoma | 44 – 72% |
| **Renal cysts** | **59 – 63%** |
| Retinal hemangioblastoma | 45 – 59% |
| **Renal cell carcinoma** | **24 – 45%** |
| Spinal cord hemangioblastoma | 13 – 59% |
| **Pheochromocytoma** | **0 – 60%** |
| Neuroendocrine tumor of pancreas | 5 – 17% |
| Serous cystadenoma of pancreas | 12 % |
| Medullary hemangioblastoma | 5 % |
| Papillary cystadenoma of epididymis | 10 – 60% |

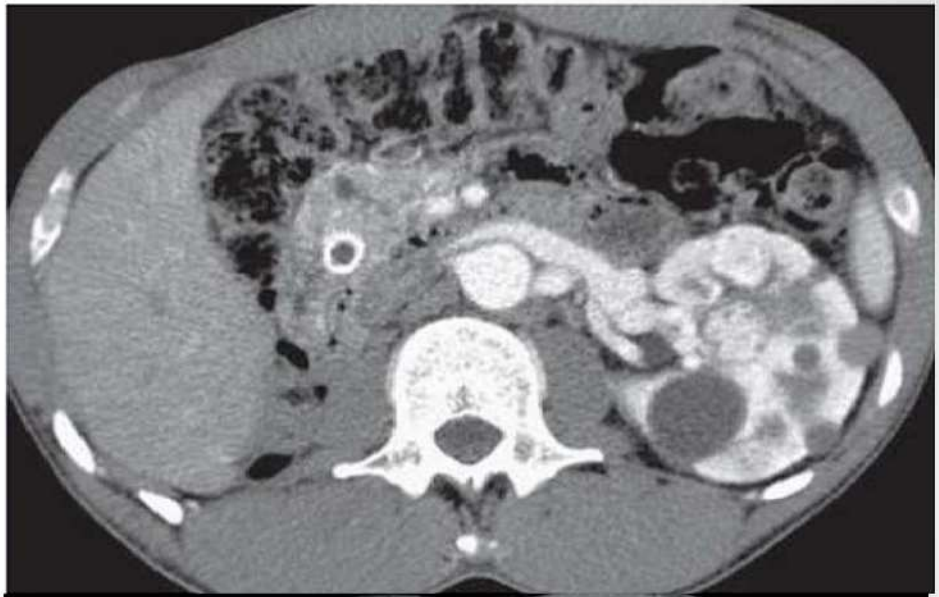


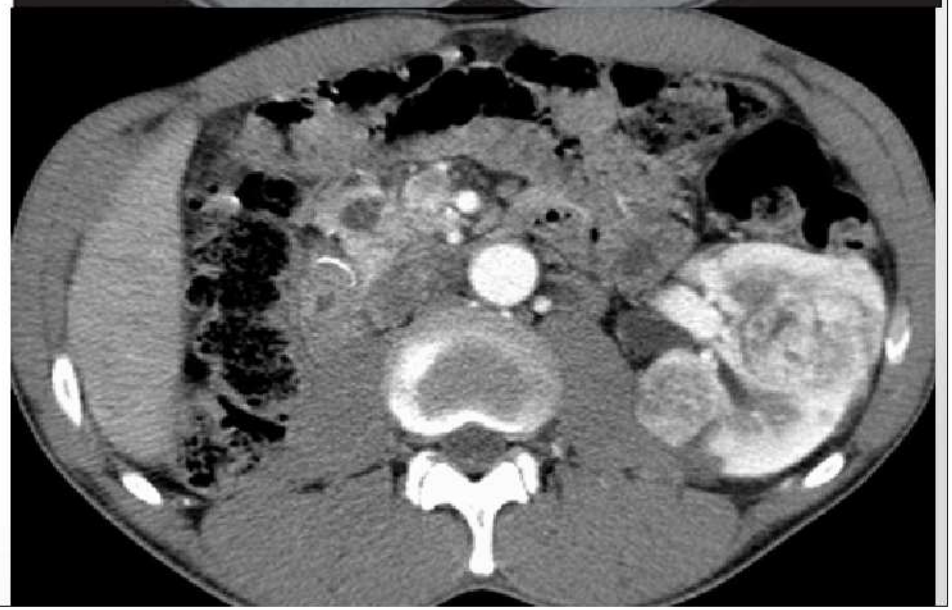

Leung RS et al. RadioGraphics 2008 : 65 - 79

# VHL




Mitmed eri segaehoogeensusega kolded – Mitmed neerurakulise vähi kolded





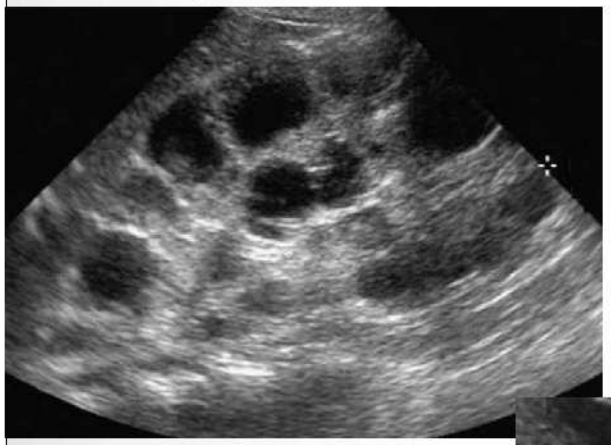
Parem neer eemaldatud, mitmed kontraseeruvad kolded vasakus neerus- RCC

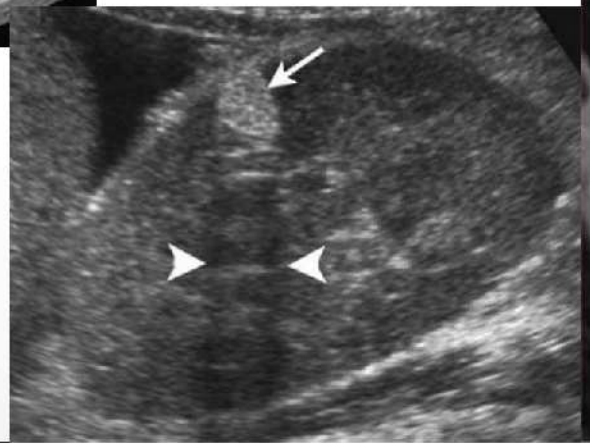
# VHL uurimisalgoritm

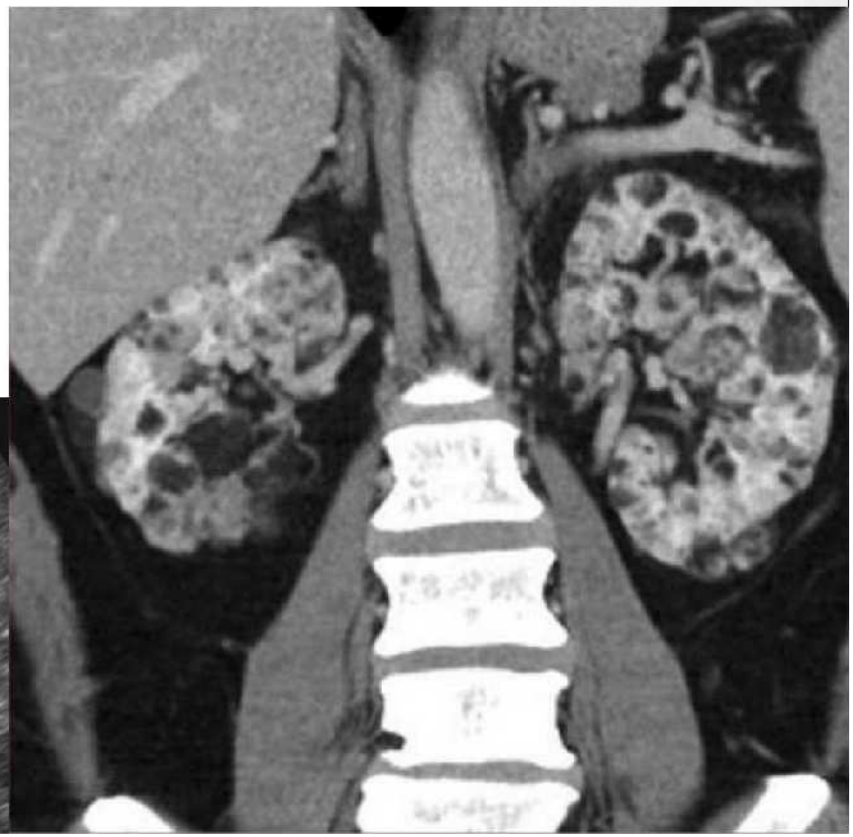
| Body System | Regimen | Follow-up |
|---|---|---|
| Renal | Annual abdominal US study from the age of 10 y | CT or MR imaging (depending on the US findings) |
| CNS | Baseline MR imaging of the brain and spine at the age of 20 y or if the patient is symptomatic; annual neurologic examination | Low threshold for repeat imaging if there are any suspicious signs or symptoms |
| Adrenal | Annual 24-h measurement of the urinary VMA level from the age of 10 y; annual blood pressure measurement | No imaging is warranted unless the urinary VMA level is abnormal |
| Ophthalmic | Annual direct and indirect ophthalmoscopy from the age of 5 y with or without fluorescein angiography | No radiologic input is required |
| Auditory | Completion of a questionnaire; an audiogram is obtained if the answers to the questionnaire are positive | If the audiogram shows abnormal results, MR imaging is performed |

# Tuberoosne skleroos

- Autosoom-dominante haigus
- Iseloomulik angiomüülipoomide teke nahas, ajus, neerudes ja teistes organites.
- Neerudes angiomüülipoom 50-70% ning lihtsad tsüstid 30-50% juhtudest



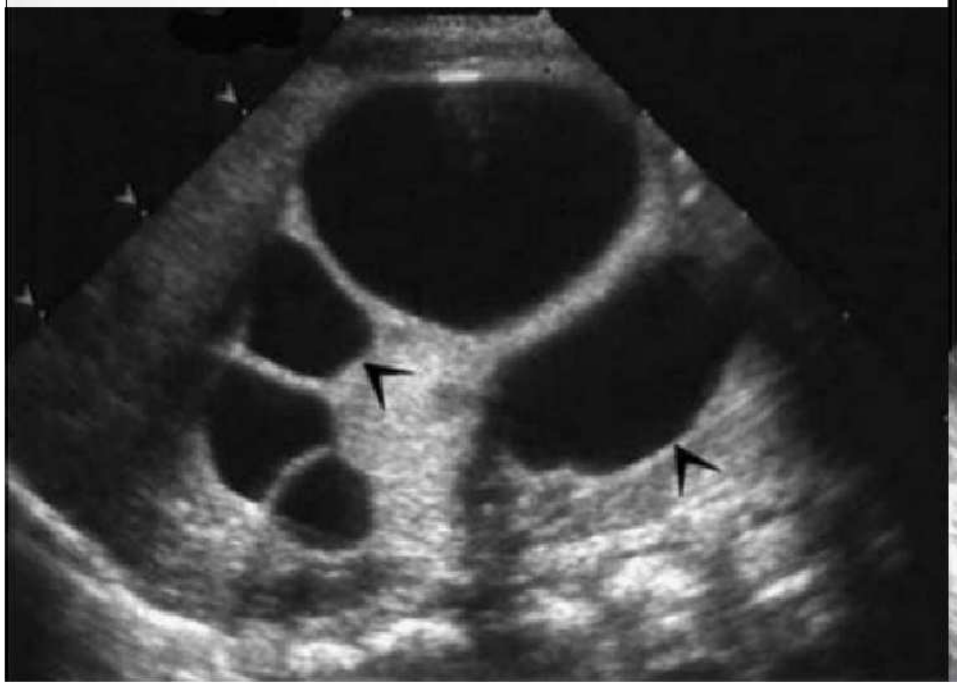


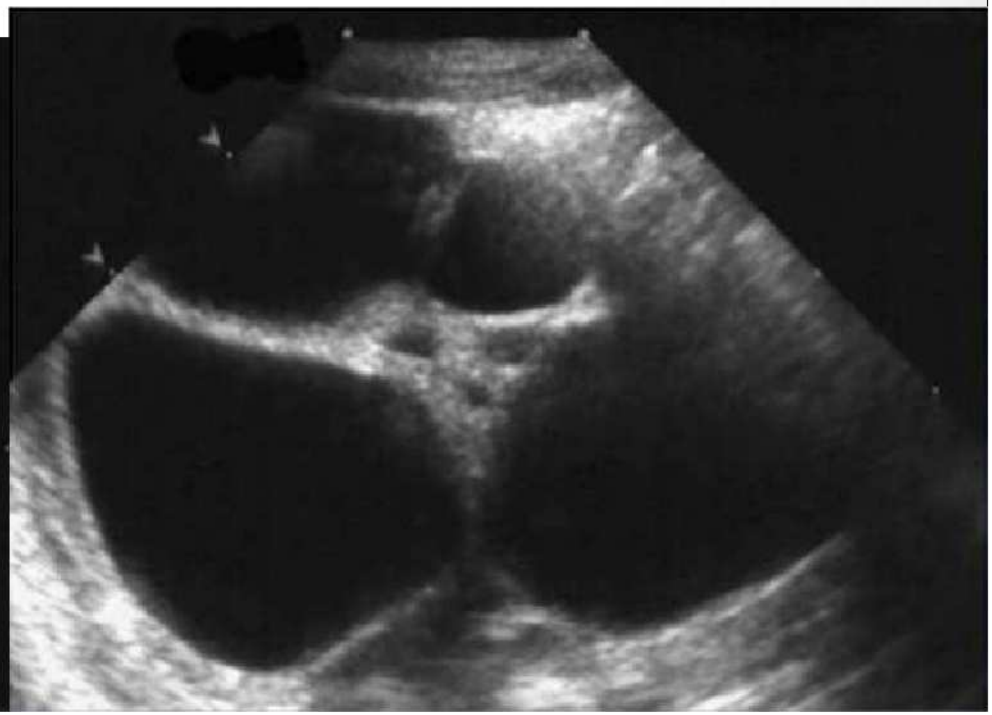


# Multitsüstilinie düsplastiline neeruhaigus

- Kaasasündinud neerude malformatsioon
- Kogu kooreosa asendunud tsüstidega

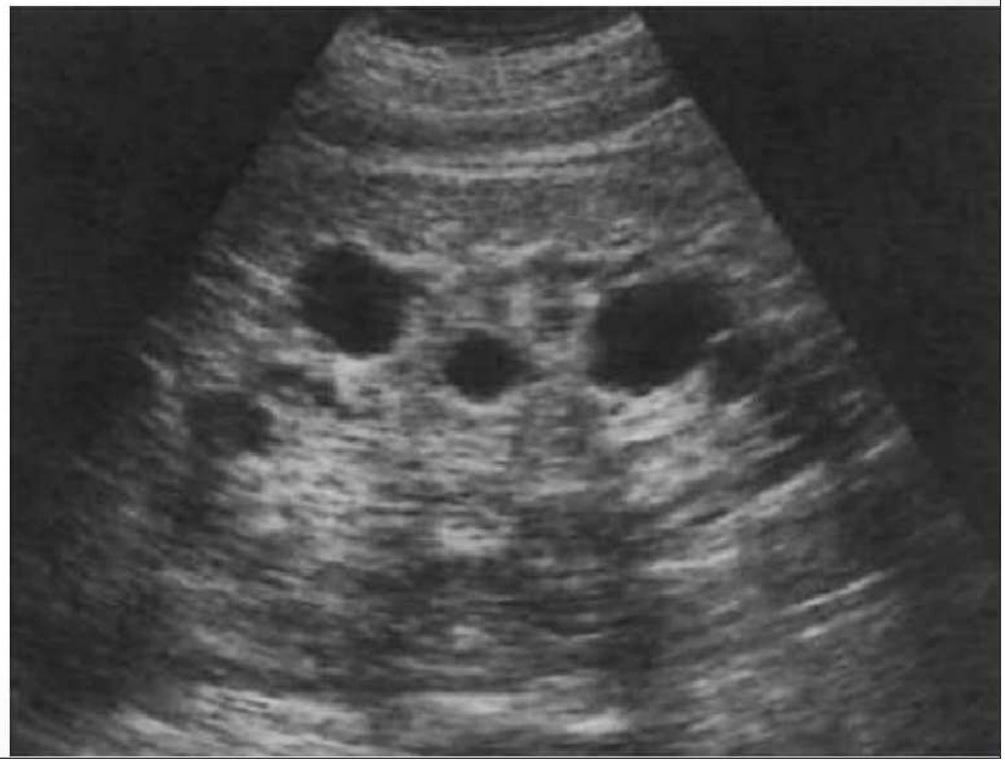
- Puudub eristatav kortikaalne parenhüüm ja tsentraalse siinuse struktuur





# Neerude omandatud tsüstiline haigus

- Tekib pikaaegsel dialüüsravi korral (lõppstaadium neerupuudulikkus)
- Sagedus tõuseb dialüüsravi pikenemisega
- Tõuseb neerurakulise vähi riski



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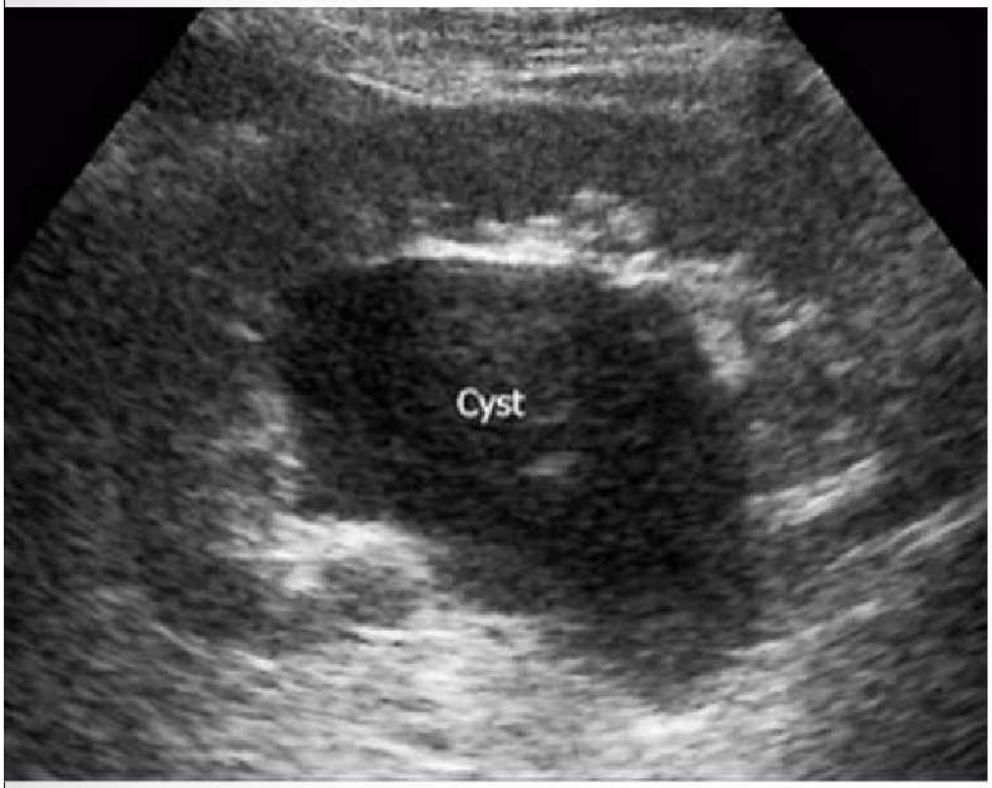
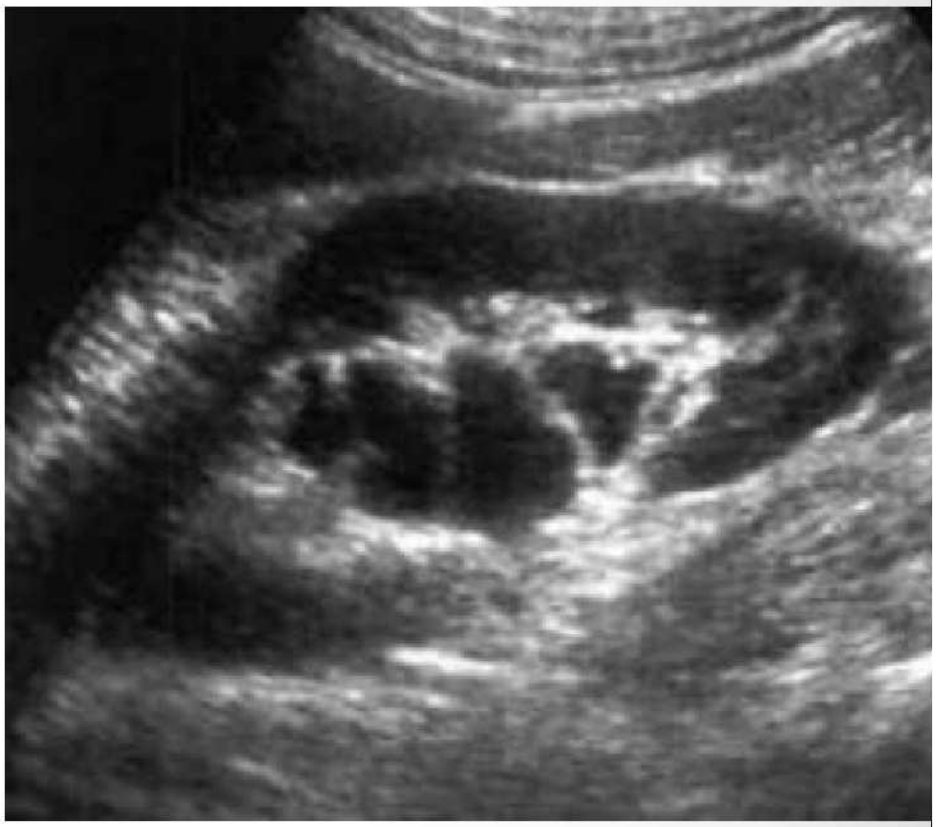
# Neeru siinuse tsüst(id)

- Parapelvikaalne: lähtub ümbritsevast parenhüümist ning võlvub neeru siinusesse. Enamasti ühepoolne ning tavalise kortikaalse tsüsti morfoloogiaga. Võib põhjustada neeruvaagna kompressiooni.
- Peripelvikaalne: healoomuline, lähtub siinusest ja omab lümfaatilist päritolu. Tavaliselt bilateraalne ning ei põhjusta kompressiooni.
- Tavaliselt ei oma kumbki kliinilist tähendust – soovitatav kasutada mõlema puhul terminit neeru siinuse tsüst (renal sinus cyst)

Rha SE et al. RadioGraphics 2004 : 2004 : 24: S117 – S131




Üksik siinuse tsüst



Mitmed siinuse tsüstid

| | | | | |
|---|---|---|---|---|
| Nephronophthisis | Small | 1MM-2CM | Medullary | Normal |
| Acquired Cysts | Normal/Small | 0.5-3CM | Any | Normal |
| Medullary Sponge | Normal/Enlarged | MM | Precalyceal | Normal |
| ARPKD | Enlarged | MM | Any | Fibrosis |
| Multicystic Dysplastic | Enlarged | 1MM-10CM | Any | Normal |
| ADPKD | Enlarged | MM-10CM | Any | Cysts |

# Kasutatud kirjandus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- Gabr, A.H., Gdor, Y., Roberts, W.W. & Wolf, J.S., 2009, **Radiographic surveillance of minimally and moderately complex renal cysts**, *BJU international*, 103(8), pp. 1116-9