

Abordagem de lesões quísticas renais na infância

E. Rosado, D. Penha, P. Cabral , P. João, A. Tavares

Serviço de Imagiologia do Hospital Prof. Dr. Fernando da Fonseca

Directora de Serviço: Dra. Manuela Baptista



Congresso Nacional de Radiologia
Vilamoura, 9-12 de Maio de 2012

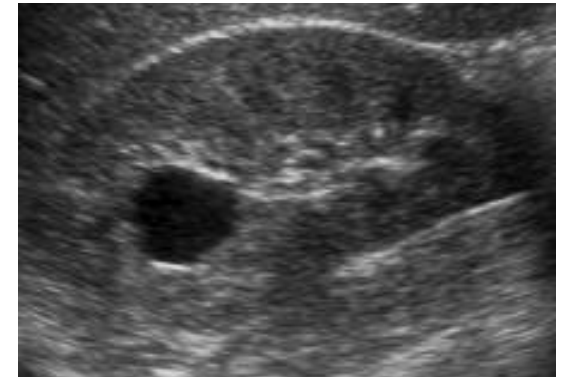
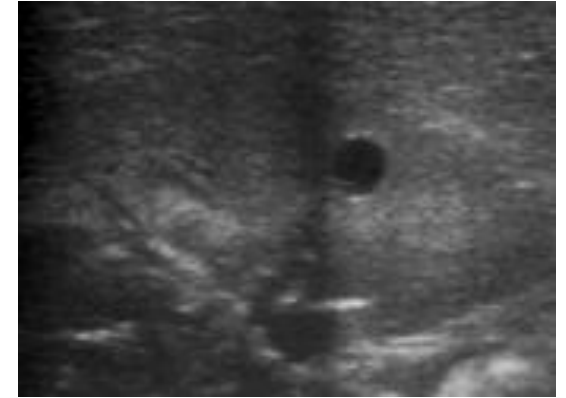
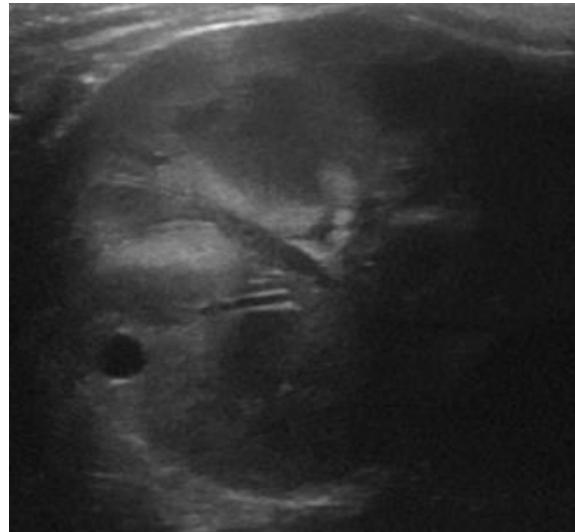
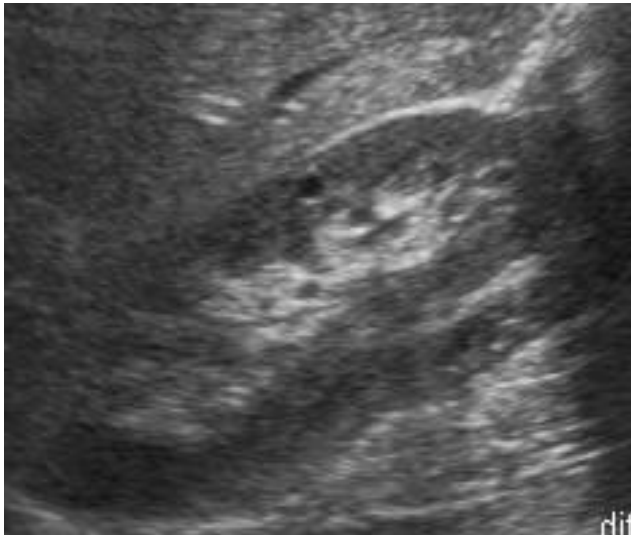


Estrutura

- Introdução
- Classificação
- Causas mais frequentes de quistos renais
 - DRPQ-AD
 - DRPQ-AR
 - Doença quística associada a doenças sistémicas
 - Rim displásico multiquístico
 - Quisto simples
- Conclusão

Introdução

- Os quistos renais nas crianças são raros



- Procurar eventual doença subjacente

Introdução

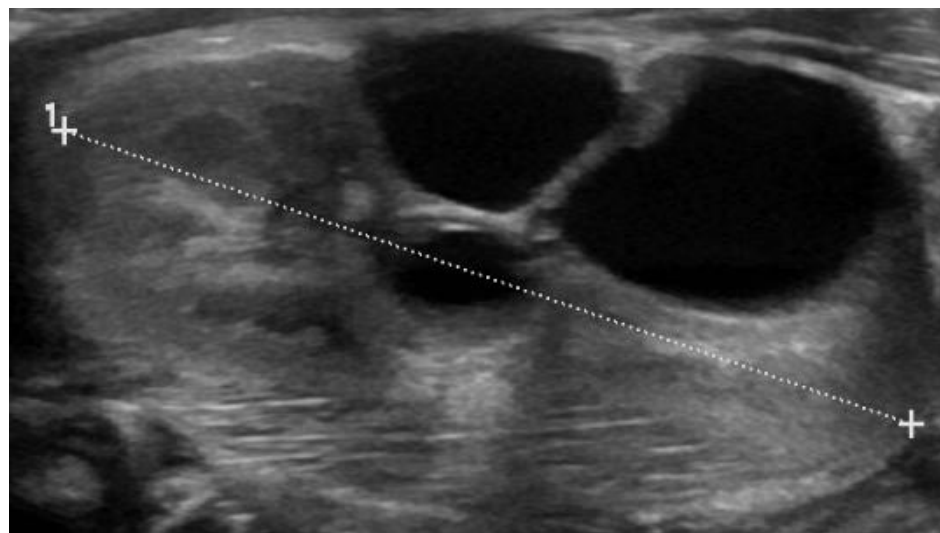
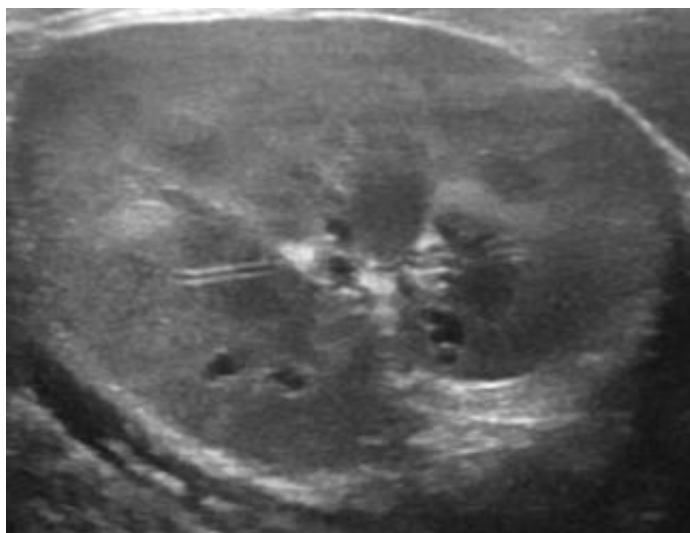
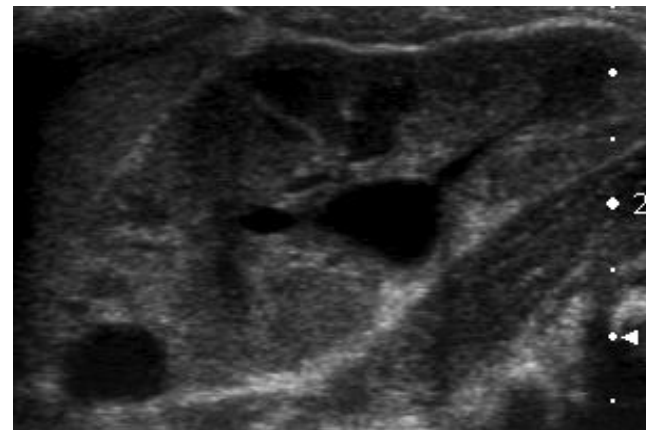
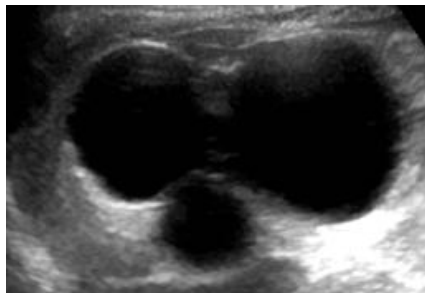
O diagnóstico depende de múltiplos factores:

- História obstétrica e ecografia pré-natal
- Antecedentes familiares
- Clínica e exame objectivo
- Exames laboratoriais
- Exames imagiológicos
- Estudo anátomo-patológico (biópsia renal)
- Estudo genético

Introdução

Métodos imagiológicos:

- Ecografia (++)
- TC
- RM (>S)



Classificação

Doenças renais quísticas na infância e adolescência

Hereditárias:

- DRPQ-AD
- DRPQ-AR
- Doença quística associada doenças sistémicas
- Doenças glomeruloquísticas
- Nefronoftise e Rim esponjoso medular
- Displasia quística hereditária
- Síndromes associadas a displasia quística medular:

Adquiridas:

- Rim displásico multiquístico
- Quistos simples idiopático
- Tumores quísticos
- Displasia quística obstrutiva

Ivemark
Bardet-Biedl
Hajdu-Cheney
Besck-with- Wiedeman
Jeune e outras Condrodisplasias
Cerebro-hepato-renal
Oro-facial-digital tipo 1
Meckel-Gruber
Trissomias 9 e 13

Classificação

Hereditárias:

- DRPQ-AD
- DRPQ-AR
- Doença quística associada a doenças sistémicas:
 - Esclerose tuberosa
 - Doença de Von-Hippel-Lindau

Adquiridas:

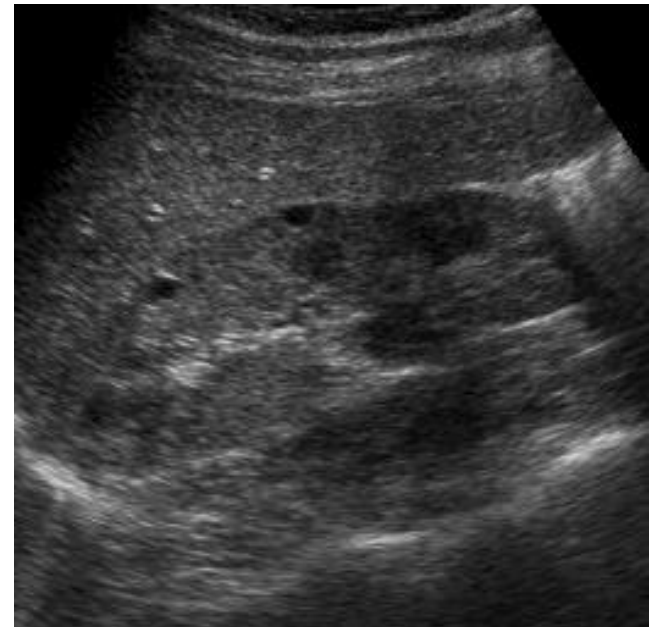
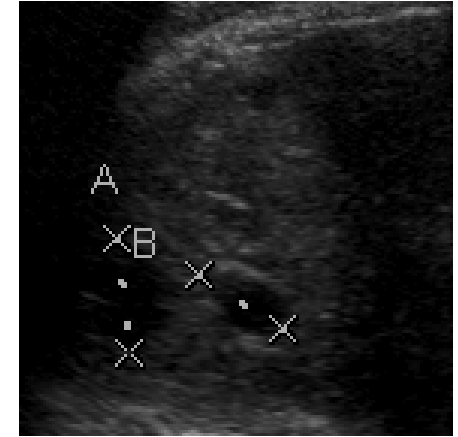
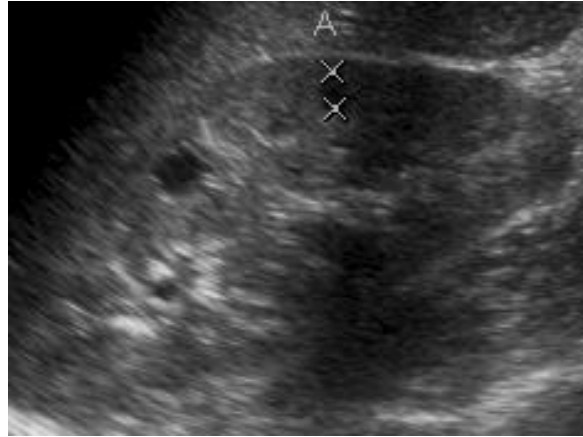
- Rim displásico multiquístico
- Quisto simples idiopático

DRPQ-AD

++ Adultos

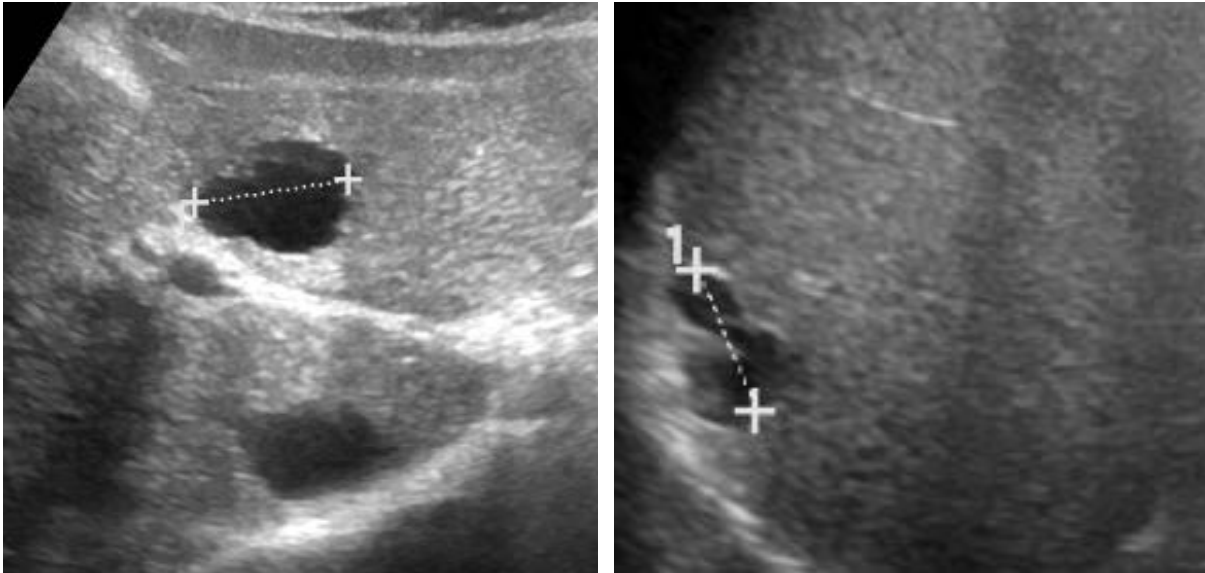
Ecografia renal:

- Volume aumentado
- Quistos corticais e medulares
- Envolvimento assimétrico
- Evolução temporal

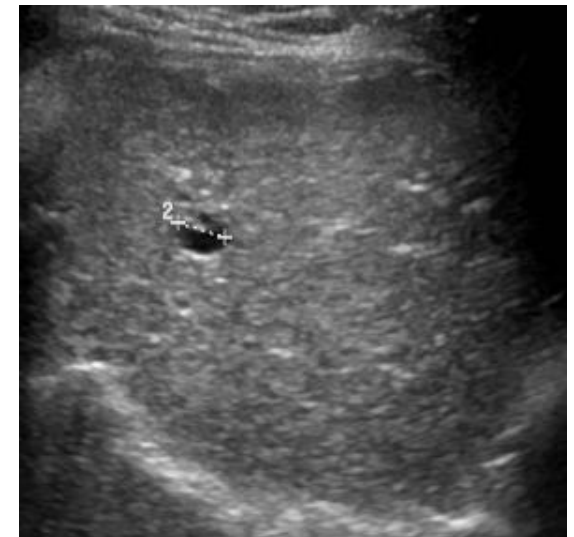


DRPQ-AD

Manifestações extra-renais:

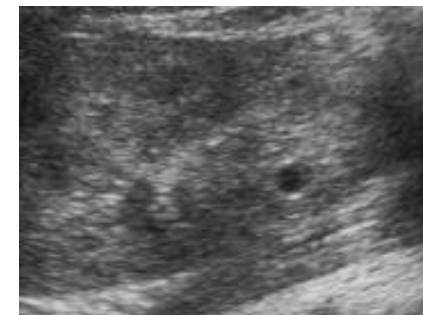
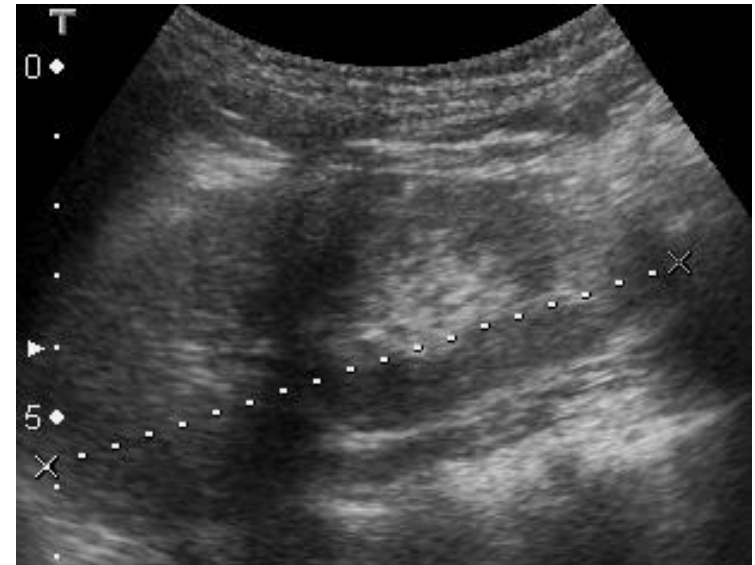
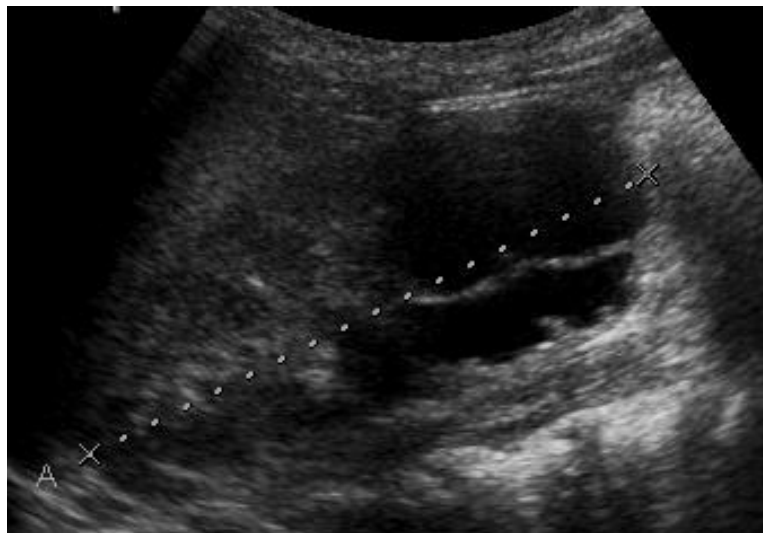


- Quistos hepáticos (++) e pancreáticos
- Aneurismas cerebrais
- Dilatação e dissecção da aorta torácica
- Prolapso da válvula mitral



DRPQ-AD

- Sexo masculino, 2000



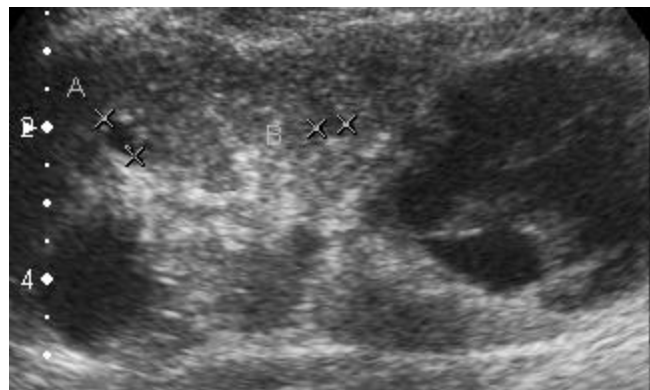
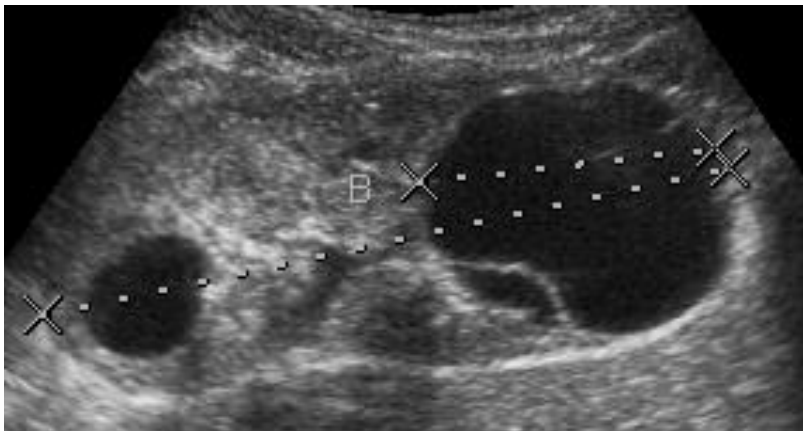
6 anos

RD: 85mm, 4 quistos

RE: 95mm, 2 quistos

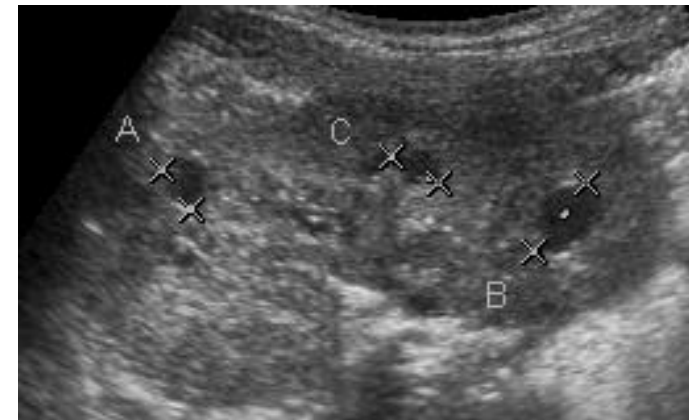
DRPQ-AD

- Sexo masculino, 2000



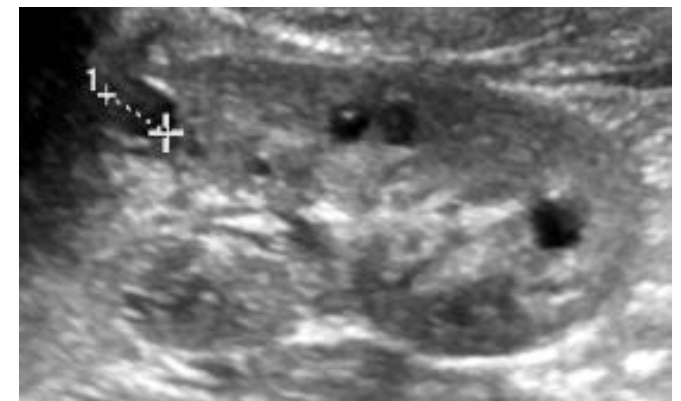
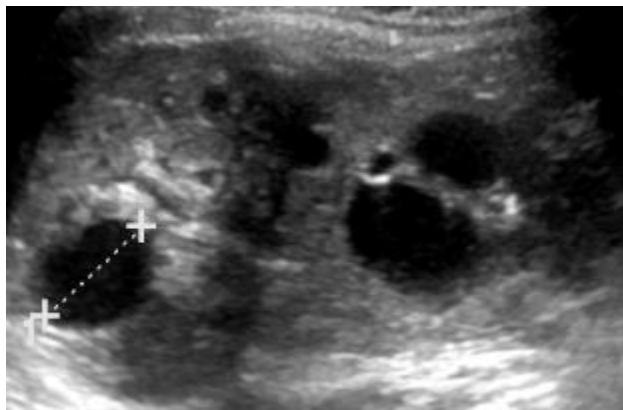
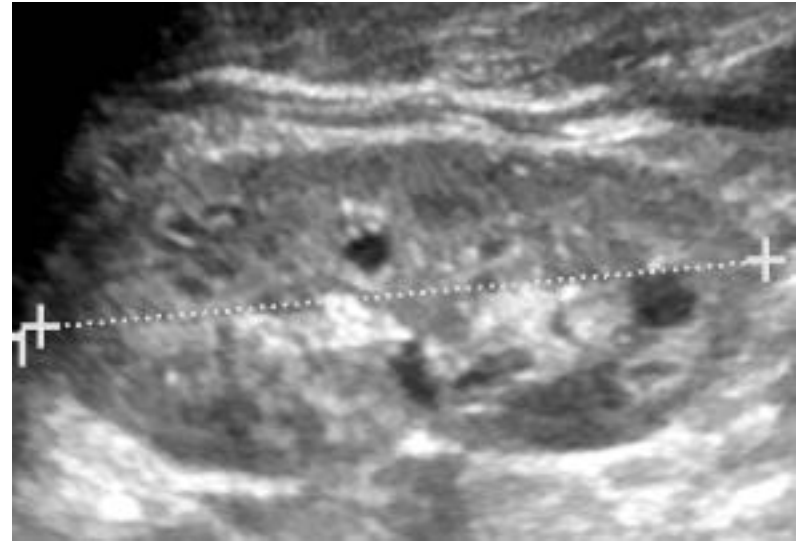
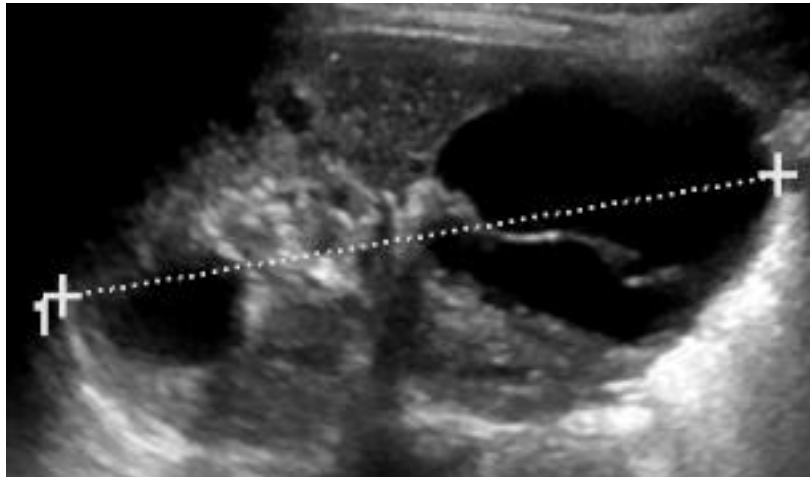
8 anos

RD: 98mm, 4 quistos
RE: 85mm, 3 quistos



DRPQ-AD

- Sexo masculino, 2000



10 anos

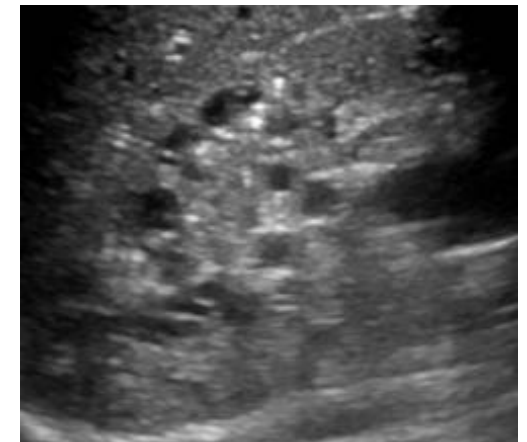
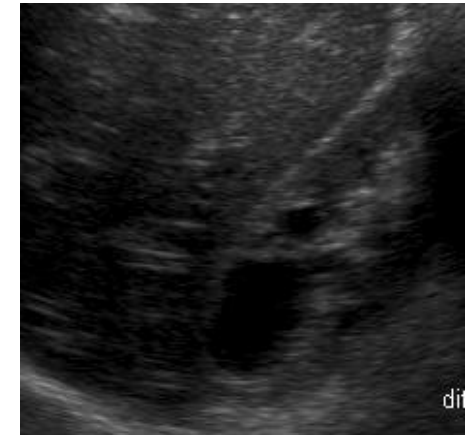
RD: 108mm, 4 quistos
RE: 90mm, 5 quistos

DRPQ-AR

- Diagnosticada in útero ou no período peri-natal

Ecografia renal:

- Volume estável
- Rins ecogénicos
- Diminuição da diferenciação
- Ductos colectores medulares dilatados
- Simétricos

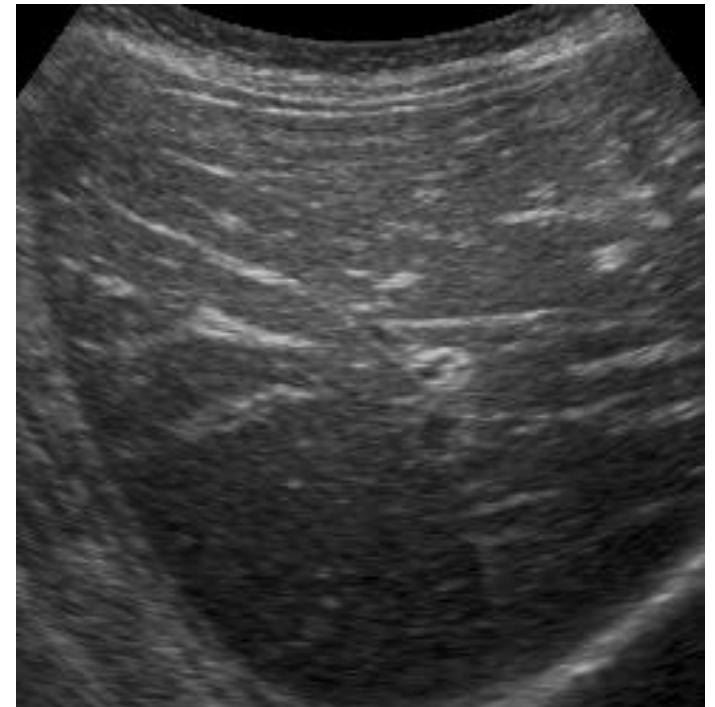
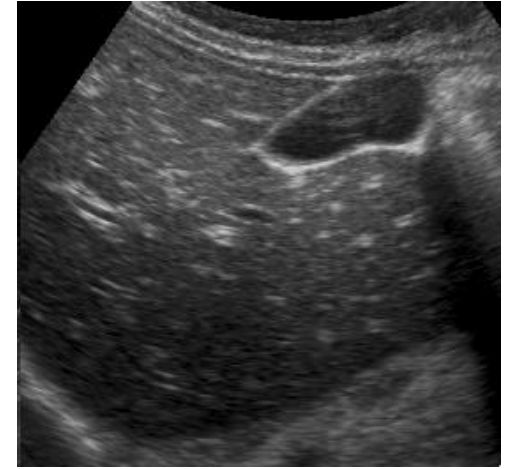


DRPQ-AR

- Doença hepatorenal fibroquística

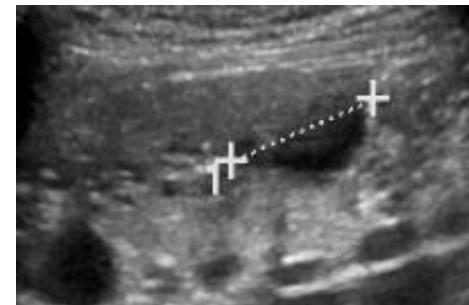
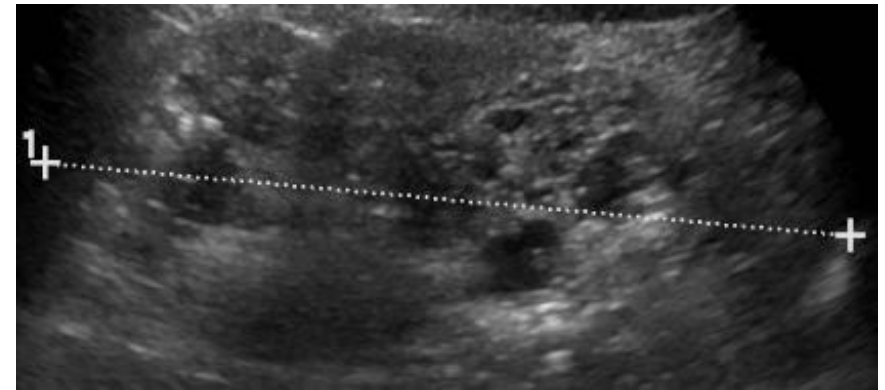
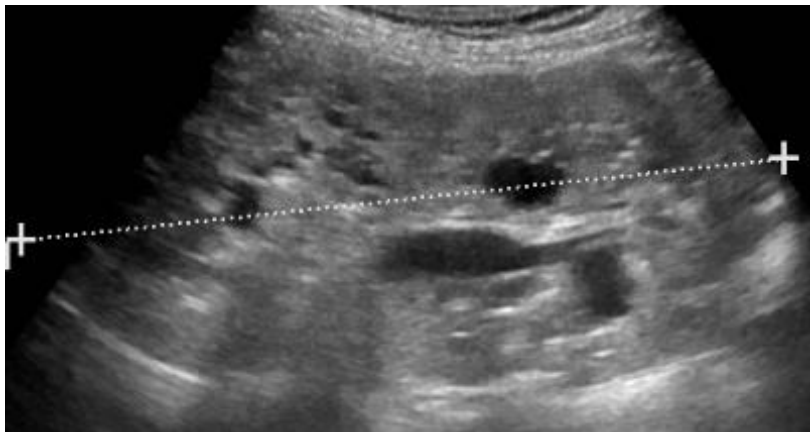
Manifestações hepáticas:

- Hepatomegália
- Hiperecogenicidade difusa
- Reforço dos espaços porta
- Dilatação dos ductos biliares
- HTP



DRPQ-AR

- Sexo feminino, 1993



RD: 149mm,
múltiplos
quistos, o
maior com
18mm

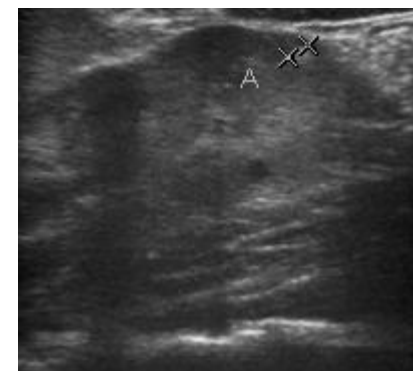
RE: 130mm,
múltiplos
quistos, o
maior com
23mm

Doenças sistêmicas

Esclerose Tuberosa

Envolvimento renal em >40% dos casos:

- Quistos simples
- Angiomiolipomas

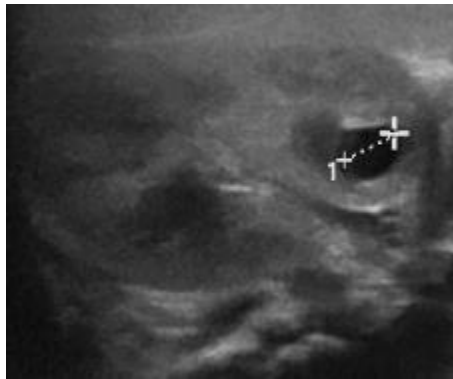


- RM crâneo-encefália
- Ecocardiograma

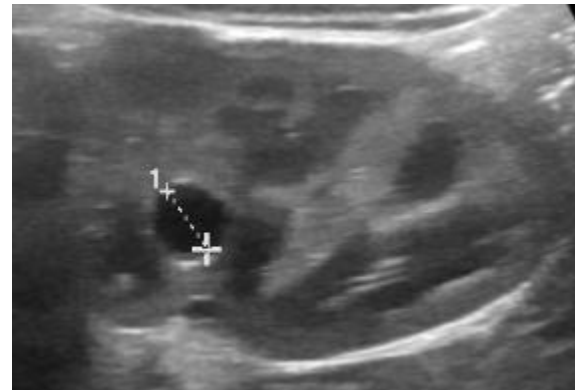
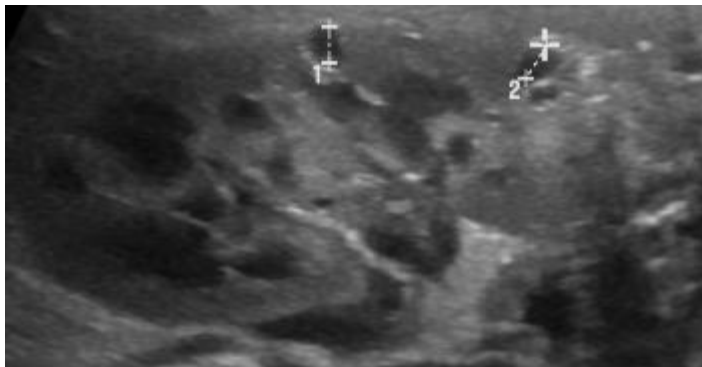
Doenças sistêmicas

- Sexo masculino, 2010

RD



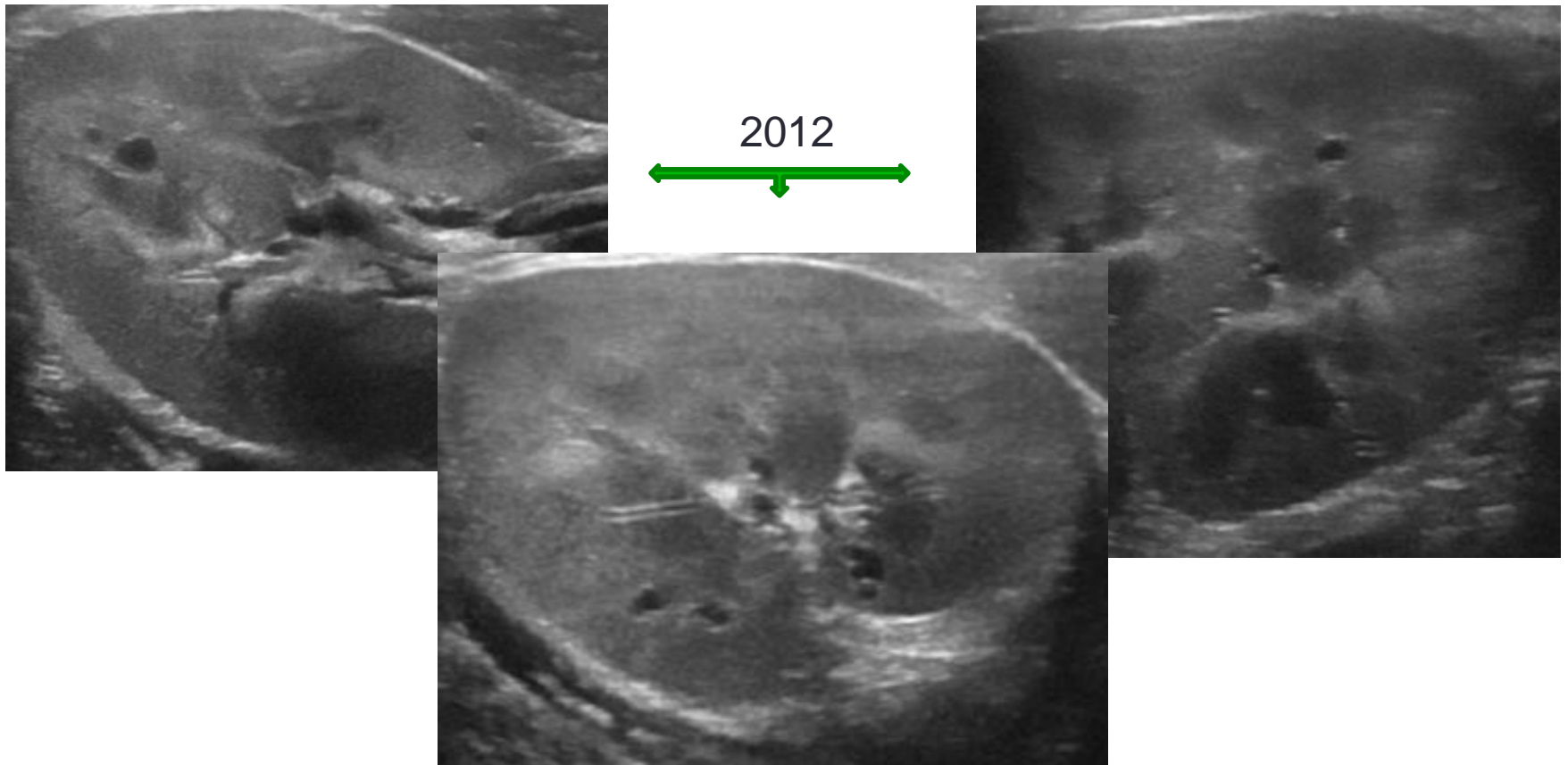
RE



2010

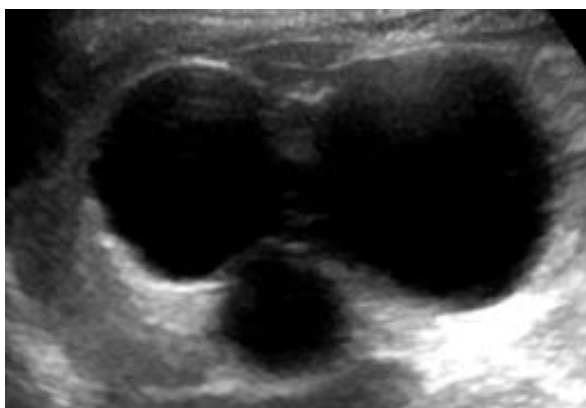
Doenças sistémicas

- Sexo masculino, 2010

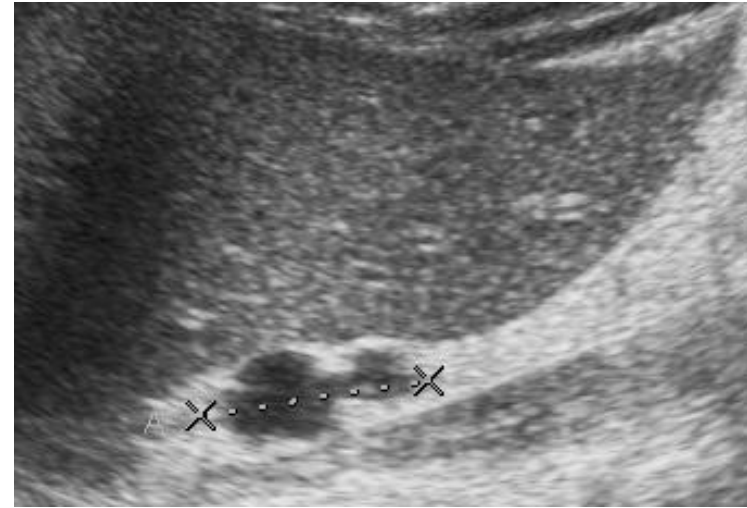
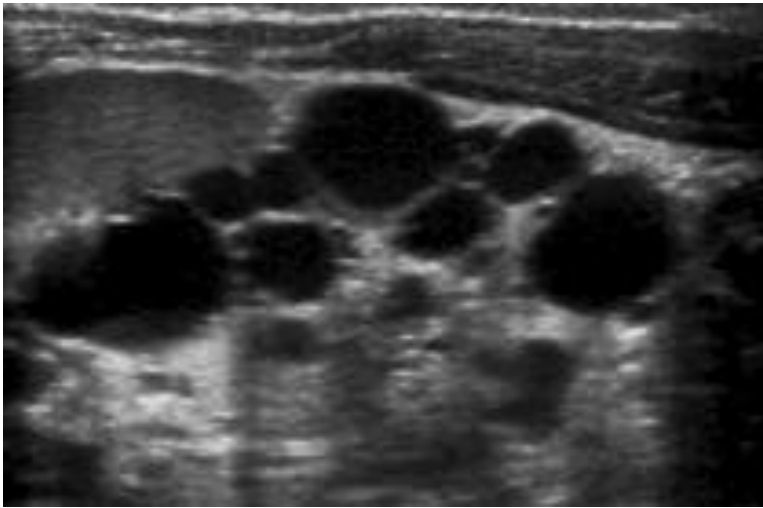


Rim displásico multiquístico

- Anomalia congénita não hereditária do desenvolvimento
- Rim não funcionante
- Unilateral
- Evolução:
 - Regressão completa
 - Calcificação
- Causa frequente de agenesia
 - Regressão completa in útero



Rim displásico multiquístico

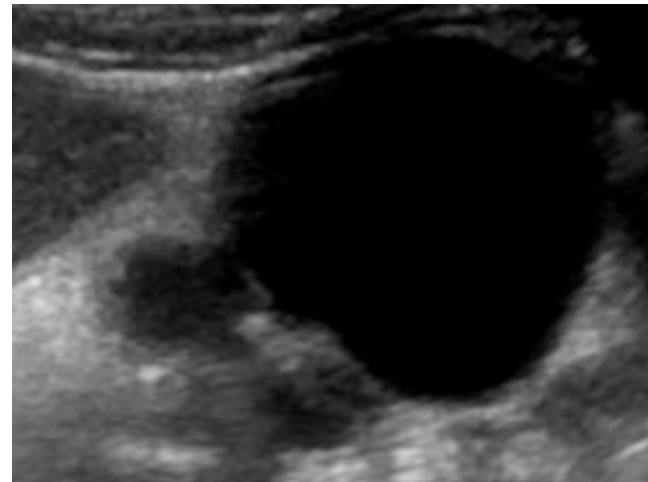


Múltiplos quistos não comunicantes

Ausência de parênquima normal

Ausência de seio renal normal

Presença de mesênquima primitivo

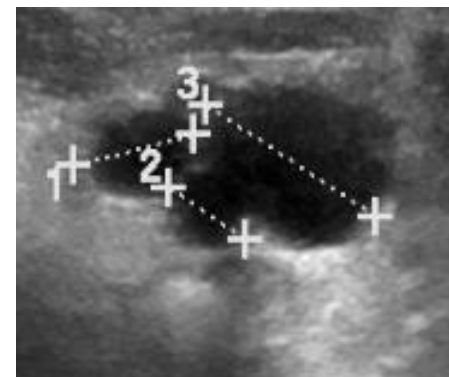
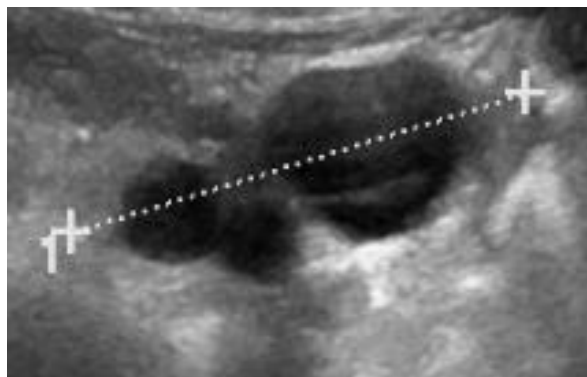


Rim displásico multiquístico

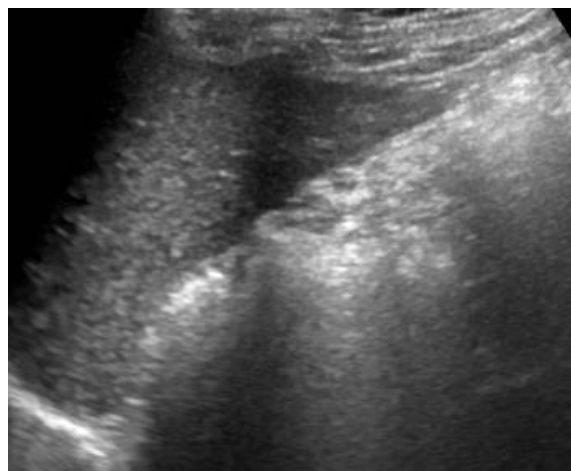
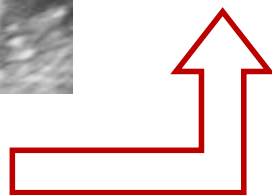
- Sexo masculino, 2009



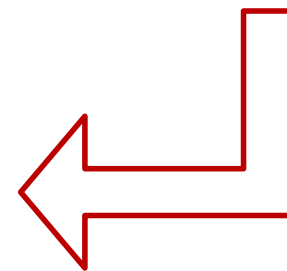
Novembro 2009



Janeiro 2011



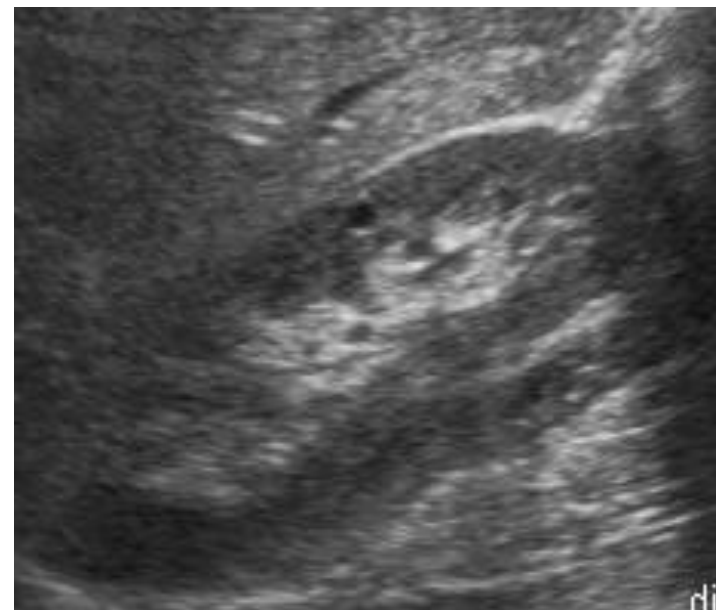
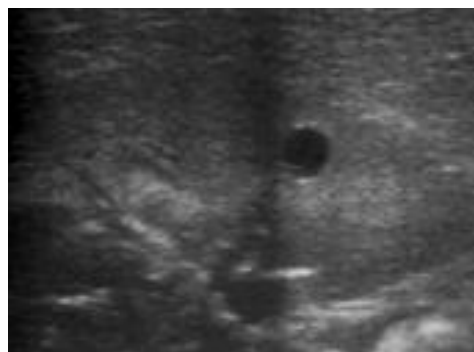
Janeiro 2012



Quisto simples idiopático

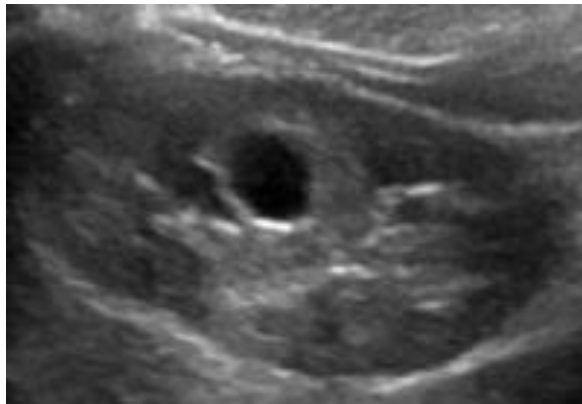
- Muito raros (frequência = 0,22%)
- Assintomáticos
- Detectados casualmente

Diagnóstico de exclusão

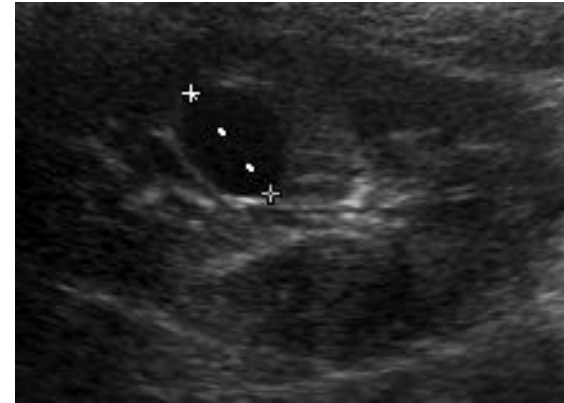


Quisto simples idiopático

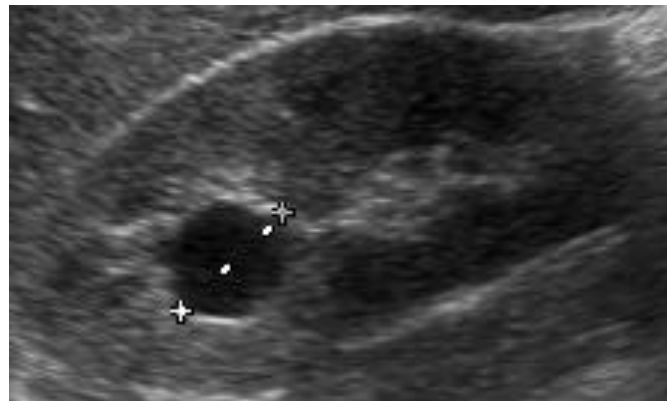
- Sexo masculino, 2006



2009: 15mm



2010: 15mm



2012: 15mm



Conclusão

- Os quistos renais nas crianças são raros
- Pesquisar doença subjacente
 - História clínica
 - Exames laboratoriais e genéticos
 - Aspecto ecográfico dos rins
 - Outros exames imagiológicos (RM)
- Manter *follow up* adequado
- Quisto simples idiopático é diagnóstico de exclusão

Bibliografia:

- Lonergan G, Col L, Rice R, Suarez E. Autosomal Recessive Polycystic Kidney disease: Radiologic-pathologic correlation. *Radiographics* 2000, 20:837-855
- Sweeney WE, Avner DE. Diagnosis and management of childhood polycystic kidney disease. *Pediatr Nephrol.* 2011, 26:675-692
- Avni FE, Hall M. Renal cystic diseases in children: new concepts. *Pediatr Radiol.* 2010, 40: 939-946
- Kiyak A, Yilmaz A, Turham P, Sander S, Aydin G, Aydogan G. Unilateral multicystic dysplastic kidney: single center experience. *Pediatr Nephrol.* 2009; 24: 99-104
- Avner DE, Sweeney WE. Renal cystic disease: new insights for the clinician. *Pediatr Clin North Am.* 2006; 53: 889-990
- Casper KA, Donnelly LF, Chen B, Bissler JJ. Tuberous sclerosis complex: renal imaging findings. *Radiology.* 2002; 225(2):451-6
- Avni FE, Guissard G, Hall M, Janssen F, DeMaertelaer V, Rypens F. Hereditary polycystic kidney diseases in children: changing sonographic patterns through childhood. *Pediatr Radiol.* 2002; 32:169-174.

Abordagem de lesões quísticas renais na infância

E. Rosado, D. Penha, P. Cabral , P. João, A. Tavares

Serviço de Imagiologia do Hospital Prof. Dr. Fernando da Fonseca

Directora de Serviço: Dra. Manuela Baptista



Congresso Nacional de Radiologia
Vilamoura, 9-12 de Maio de 2012

