

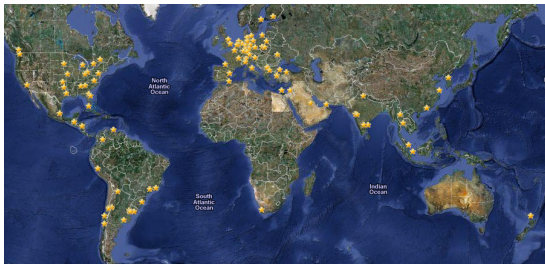


PD in Special Pediatric Populations

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The International Pediatric PD Network



Registered Network users: 243 centers in 51 countries
Registry participants: 127 centers in 43 countries
3,648 patients enrolled as of March 15, 2019

Pediatr Nephrol (2013) 28:315–319
DOI 10.1007/s00467-012-2303-9

ORIGINAL ARTICLE

Ventriculoperitoneal shunts in children on peritoneal dialysis: a survey of the International Pediatric Peritoneal Dialysis Network

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VP Shunts in Children Undergoing Chronic PD

- Survey answered by 76 IPPN sites
- **18 patients** at 13 sites concomitant VPS and PD caths (VPS first in 15)
- Global prevalence: 1/153 children
- Mean PD duration with VPS in place: 23 months
- 20 peritonitis episodes (11 pts, 392 months observation)
- No ascending shunt infection during peritonitis episode
- In 1 patient temporally unrelated peritonitis/VPS infection episodes
- No other complications of catheter dysfunction

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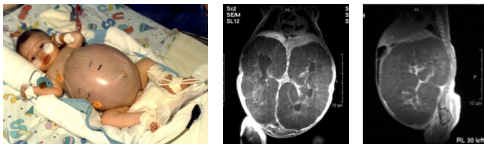
HD in an infant when PD is not possible

- Very large cystic dysplastic kidneys (ARPKD)
- Intra-abdominal pathology
 - necrotising enterocolitis / inflammatory bowel pathology
 - after abdominal surgery
 - abdominal wall defects - gastroschisis / bladder extrophy
 - anorectal malformations
 - (colostomy)

c/o R.Shroff

'Malignant' Neonatal Form of ARPKD

Admitted for hypertension, dehydration.
BP 222/133 mm Hg, LVH. Scr: 0.76 mg/dl



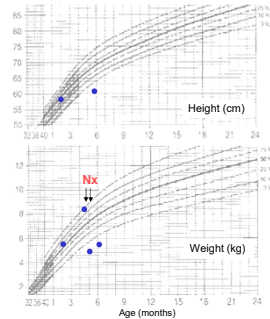
Compression of inferior vena cava, enteral nutrition impossible.
Rapid loss of renal function, oliguria

Emergency unilateral Nx, followed by contralateral Nx,
Tenckhoff catheter placement, start of PD

Malignant Neonatal Form of ARPKD



- 3.3 kg kidney



Chronic PD in Children with ARPKD

Background

- Potential limitation: insufficient intraabdominal space for PD due to nephro/hepato/splenomegaly
- Neonatal form: guarded prognosis of pulmonary hypodysplasia; aggravated by PD?
- Leukopenia due to splenomegaly -> infection risks?
- Post-nephrectomy hypotension

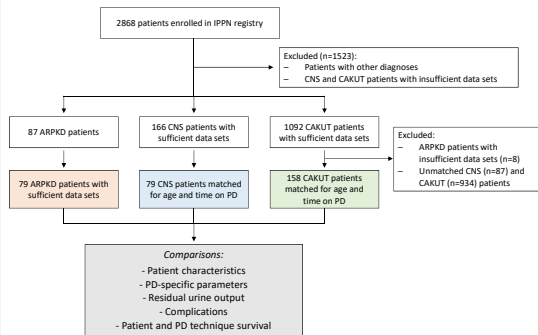
4.3% of children in IPPN Registry have ARPKD

Aim of study

- To explore PD prescription and outcomes in children with ARPKD

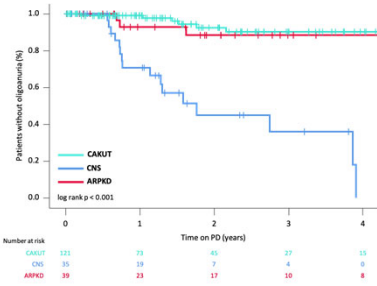
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Cohort Study with Case-Control Design



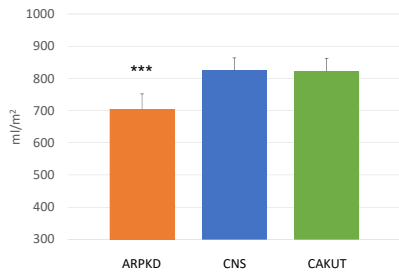
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Residual Renal Function



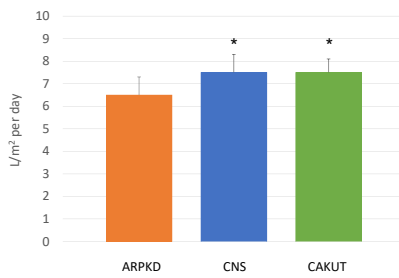
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Peritoneal Fill Volume



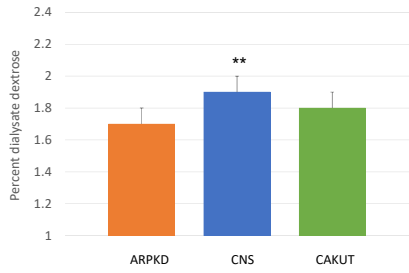
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Total PD Fluid Turnover (APD)



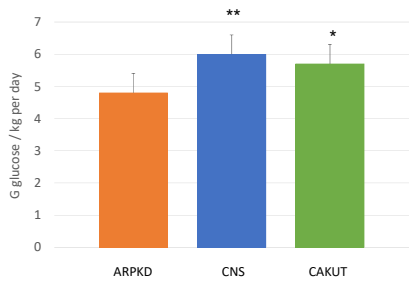
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Peritoneal Glucose Concentration



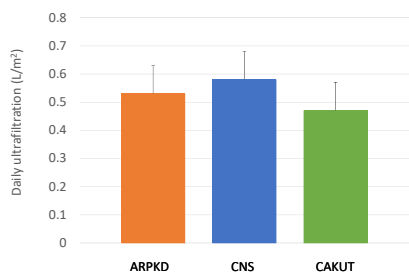
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Peritoneal Glucose Exposure



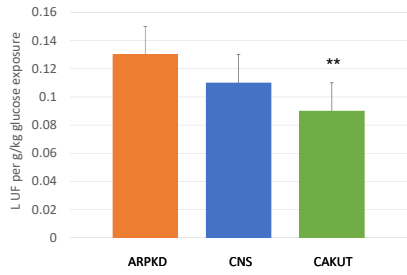
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Daily Ultrafiltration



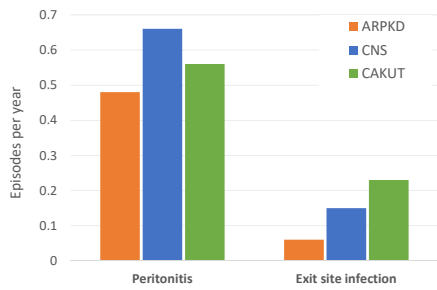
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Ultrafiltration Efficiency



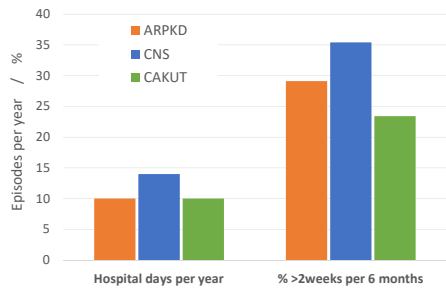
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PD-Related Infections



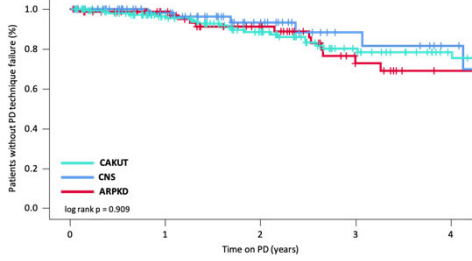
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Hospitalization Rates



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PD Technique Survival



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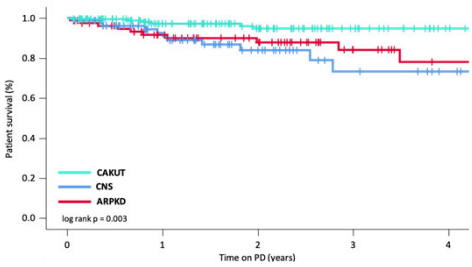
Risk Factors for Technique Failure

Cox proportional hazard analysis

Variable	HR	95% CI	P-value
Diagnosis			
ARPKD (N=72) (reference)	1.000		
CNS (N=70)	0.786	0.313 – 1.970	0.607
CAKUT (N=138)	0.926	0.439 – 1.954	0.839
Oligoanuria status at baseline	1.506	0.769 – 2.951	0.232
Age in years	0.823	0.734 – 0.922	0.001
Gender	1.357	0.692 – 2.659	0.374

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Patient Survival



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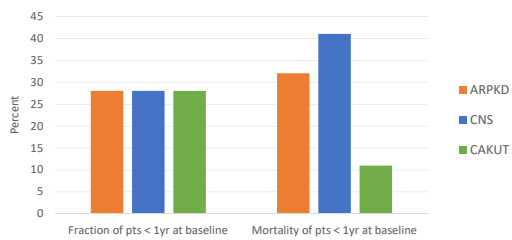
Mortality Risk Factors

Cox proportional hazard analysis

Variable	HR	95% CI	P-value
Diagnosis			
ARPKD (N=72)	1.000		
CNS (N=70)	1.416	0.616 – 3.254	0.412
CAKUT (N=138)	0.291	0.103 – 0.824	0.020
Oligoanuria status at baseline	1.301	0.551 – 3.076	0.548
Age in years	0.413	0.285 – 0.598	<0.001
Gender	0.871	0.401 – 1.891	0.727

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Infant Mortality



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Causes of Death

ARPKD patients (n=13)

- 5 non-PD-related infections
- 3 cardiac (sudden arrest, cardiorespiratory failure, "heart attack")
- 2 non-infectious PD-related complications
- 1 brain calcifications and severe hyperlipidemia
- 1 seizures with cardiac arrest
- 1 unknown

CNS patients (n=13)

- 5 pulmonary edema / congestive heart failure

Non-PD related infections tended to be more common causes of death in ARPKD (p=0.12)

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HD in an infant when PD is not possible

- Very large cystic dysplastic kidneys (ARPKD)
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 - after abdominal surgery
 - abdominal wall defects - gastroschisis / bladder extrophy
 - anorectal malformations
 - (colostomy)

c/o R.Shroff

PD in Children with Colostomies

Of 135 centers participating in the IPPN, 15 centers reported **20 children** on chronic PD with co-existing colostomies

Renal diagnoses: CAKUT (n=16, 80%)

Main indications for colostomy:

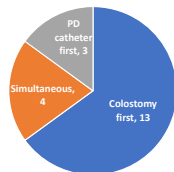
- 7, Anorectal malformation
- 4, VACTERL association
- 3, Cloacal malformation
- 3, Necrotizing enterocolitis
- 3, other

Median **age at colostomy** creation: 0.1 [IQR 0-2.2] months
Median **age at PD initiation:** 2.8 [IQR 0.2-19] months

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PD in Children with Colostomies

Sequence of Placements



Observation time with colostomy and PD cath in place together: **18** [IQR 5-36] months

Other ostomies *in situ*:

3 gastrostomies, 3 vesicostomies, 1 both gastrostomy and vesicostomy

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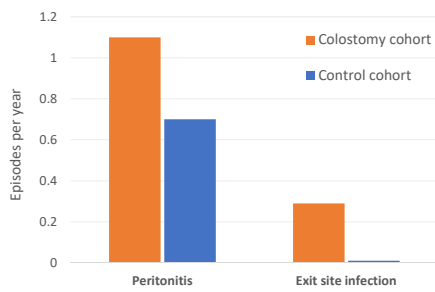
Risk of PD-Related Infection

	Colostomy	Matched* Controls
N	20	46
Time at risk (total months)	413	977
Time at risk (months per patient)	21	21
Age at PD start (months)	3 (0-18)	7 (3-11)
% pts with other -ostomies	9 (45%)	22 (48%)

* Matched by age, observation time underlying renal disease, center

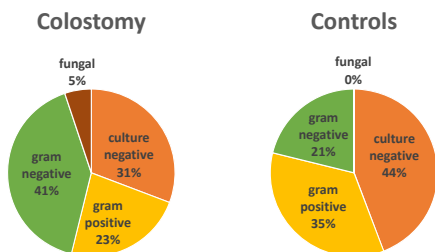
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Risk of PD-Related Infection



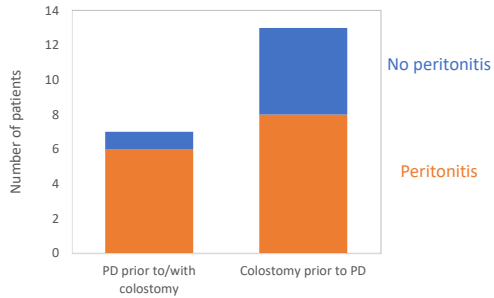
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Distribution of Causative Organisms



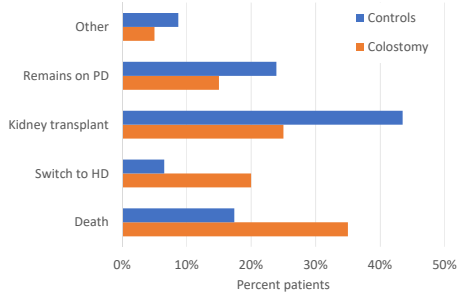
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Timing of Colostomy Placement and Peritonitis Risk



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Patient Outcomes



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Summary

- In principle, chronic PD is possible in patients with
 - indwelling VP catheters
 - ARPKD
 - colostomies

Children with ARPKD:

- Fill volumes, daily PD fluid turnover reduced by 15%
- Lower peritoneal glucose exposure required to achieve similar UF
-> non-osmotic drainage of ascites due to portal hypertension?
- Comparable complication rates, PD technique survival as age-matched other populations
- Increased mortality risk of young infants with ARPKD, comparable to CNS

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Summary

PD in children with colostomies:

- Increased risk of death
- Increased risk of PD technique failure
- 70% increased risk of (gramnegative) peritonitis
- Increased peritonitis risk if colostomy placed with or after PD catheter
- Increased risk of exit site infections

➤ Despite increased complication risks,
majority of ARPKD and colostomy patients achieve satisfactory outcomes
