

# The Dutch initiative: The EPS registry

4<sup>th</sup> February 2010

Mario Korte

- Incidence
- Dutch Multicenter Study
- EPS registry
- Aims & Challenges
- Design
- Website
- Update registry

## Garosi and Oreopoulos:

“No Need for an expiray date in chronic peritoneal dialysis to prevent EPS”. Int Urol Nephrol 2009.

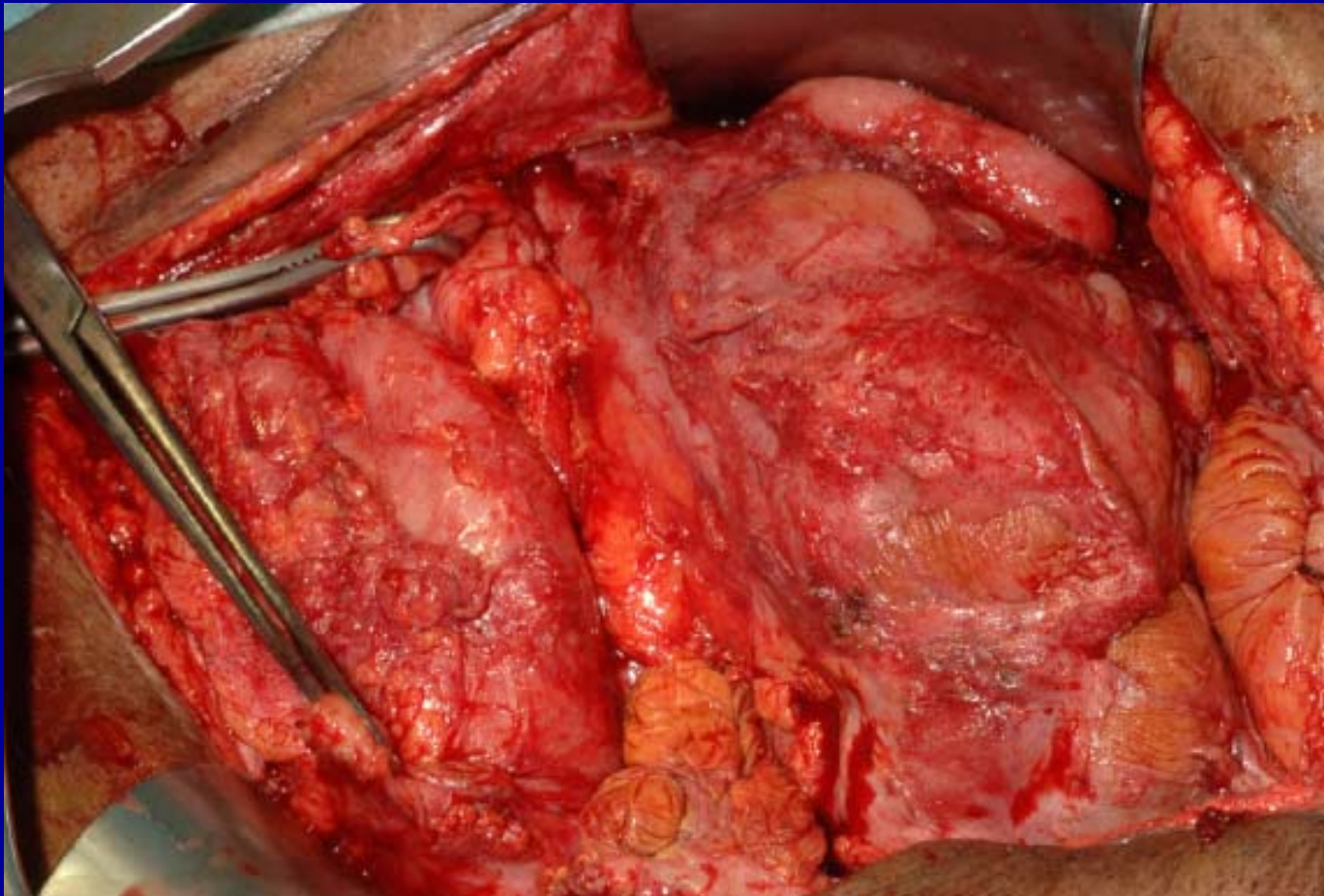
“We should concentrate on ways to decrease risk of EPS.... Many years this has been the Holy Grail of PD.”

The logo consists of a white oval shape with a blue border. Inside the oval, there is a smaller blue oval containing the white text "EPS".

EPS



Macroscopic: adhesions, fibrin depositions, thickening visceral peritoneum, cocooning



# Incidence

- Prevalence 0.7%<sup>1</sup> - 3.3%<sup>2</sup>
- Japan 1999-2003 <sup>3</sup>: incidence
  - 3 years PD 0.7%
  - 5 years PD 2.1%
  - 8 years PD 5.9%
- Netherlands:
  - 3.5 per 1000 patient years <sup>4</sup>

<sup>1</sup>Rigby Nephrol Dial Trans 1998, <sup>2</sup>Summers KI 2005, <sup>3</sup>Kawanishi AJKD 2004, <sup>4</sup>Hendriks 1997 PDI.

# Increasing Incidence?

Fieren et al. PDI 2007:619-624.

# 2006: Increasing Incidence Utrecht and Rotterdam?

- 1996-2006 18 cases.
- Cluster of cases in 2004 and 2005 (12 patients).
- PD population was stable.
- Mean PD duration  $71.8 \pm 44.9$  months.
- 17 patients icodextrin (mean  $34.2 \pm 22.2$  months).
- 83 % history of kidney transplantation (mean time to EPS  $39.3 \pm 71.1$  months).

# Concerns about EPS

Topley N. *Encapsulating peritoneal sclerosis: time to act.* PDI 2006.

Summers A et al. *An international encapsulating peritoneal sclerosis registry and DNA bank: why we need one.* PDI 2006.

Fieren M et al. *Posttransplant encapsulating peritoneal sclerosis: a worrying new trend?* PDI 2007.

# Dutch initiative EPS registry

## Nationwide concerns among nephrologists

16% PD not a first choice because risk EPS.

66% wants a national registry EPS <sup>1</sup>.

<sup>1</sup> A.M. Coester, R.T. Krediet, E.W. Boeschoten. Presented at ISPD 2008



# Questions for EPS registry

- Increasing incidence of EPS?
- What riskfactors can be identified?
- Uniform management strategies possible?

# Steering committee

## Dutch initiative EPS registry

Project leaders

M.Korte (Albert Schweitzer hospital)

M.Betjes (Erasmus MC)

Hans Mak institute

E. Boeschoten

Epidemiology Erasmus MC

E. Steyerberg, H.Lingsma

Erasmus MC

M. Fieren

Utrecht MC

W. Boer

VUMC

F. Van Ittersum

UMCN

R. De Sevaux

AMC

D. Struijk

UMC Groningen

R. Westerhuis

AZM

F. v.d. Sande

The logo consists of a white oval shape with a blue horizontal bar across its center. The letters "EPS" are written in white on the blue bar.

EPS

# Aims & Opportunities

## Dutch initiative EPS registry

- Extending knowledge EPS
- Uniform definitions for diagnosis and management protocols
- Nationwide registry
- Epidemiological analyses of EPS
- Indicating future research areas
- Collaboration within Europe

# Challenges EPS registry

- Uniform EPS diagnosis
- Different stages of EPS: determination what stages included
- Complete dataset with all possible variables
- Easy accessible website and easy used

# Diagnosis Encapsulating Peritoneal Sclerosis (EPS)

- Clinical syndrome with persistent, intermittent or recurrent presence of intestinal obstruction
- With or without the existence of inflammation parameters
- Peritoneal thickening, sclerosis, calcifications and encapsulation confirmed by macroscopic inspection or radiological findings

Kawaguchi Y, et al. *Encapsulating peritoneal sclerosis: definition, etiology, diagnosis, and treatment. ISPD Ad Hoc Committee. PDI 2000; 20: S43-55*

The logo consists of a white oval with a blue border, containing a smaller blue oval with the white text "EPS" inside.

EPS

ISPD <sup>1</sup>	Criteria
EPS	Intestinal obstruction and Radiological or macroscopical evidence
<b>EPS registry</b>	
Macroscopical EPS ( <i>golden standard</i> )	Intestinal obstruction and Macroscopically identified EPS
Clinical EPS	Intestinal obstruction and Radiological EPS
Suspected early EPS	Intestinal obstruction <i>or</i> Two or more findings of: <ul style="list-style-type: none"> <li>- weight or appetite loss</li> <li>- bloody ascites</li> <li>- radiological suggestion of EPS</li> <li>- fast transport status or ultrafiltration failure</li> </ul>
No EPS	Intestinal obstruction, but Other cause than EPS identified with certainty



EPS

# Challenges EPS registry

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# Stages EPS

Stage	Pathological findings	Clinical findings
Pre-EPS period/asymptomatic	Peritoneal thickening and sclerosis Calcification of peritoneum	Ultrafiltration failure High transporter status Bloody dialysate, ascites Hypoproteinemia
Inflammation period	Inflammation Fibrin degradation products	Increase in CRP Leucocytosis Fever Bloody dialysate, ascites Weight and appetite loss Diarrhea and stool changes
Progressive or encapsulating period	Less inflammation Adhesions and continuation encapsulating	Disappearance of inflammation signs Appearance of symptoms of ileus: <ul style="list-style-type: none"> <li>- Nausea, vomiting,</li> <li>- Abdominal pain, constipation,</li> <li>- abdominal mass, ascites</li> </ul>
Complete ileus	no inflammation	Complete ileus, anorexia

Nakamoto H. Encapsulating peritoneal sclerosis--a clinician's approach. *Perit Dial Int* 2005; 25 Suppl 4: S30-38

EPS

# Challenges EPS registry

- Uniform EPS diagnosis
- Different stages of EPS: determination what stages included
- Design with complete dataset including all possible variables
- Easy accessible website and easy used

**Classic - conditioning riskfactors**

- Uremia
- Predisposition, age, renal diagn
- Desinfectant, practolol \**
- PD duration
- PD catheter
- Medication
- Peritonitis
- PDF's: Glucose, GDP/AGE's, low pH, lactate buffer

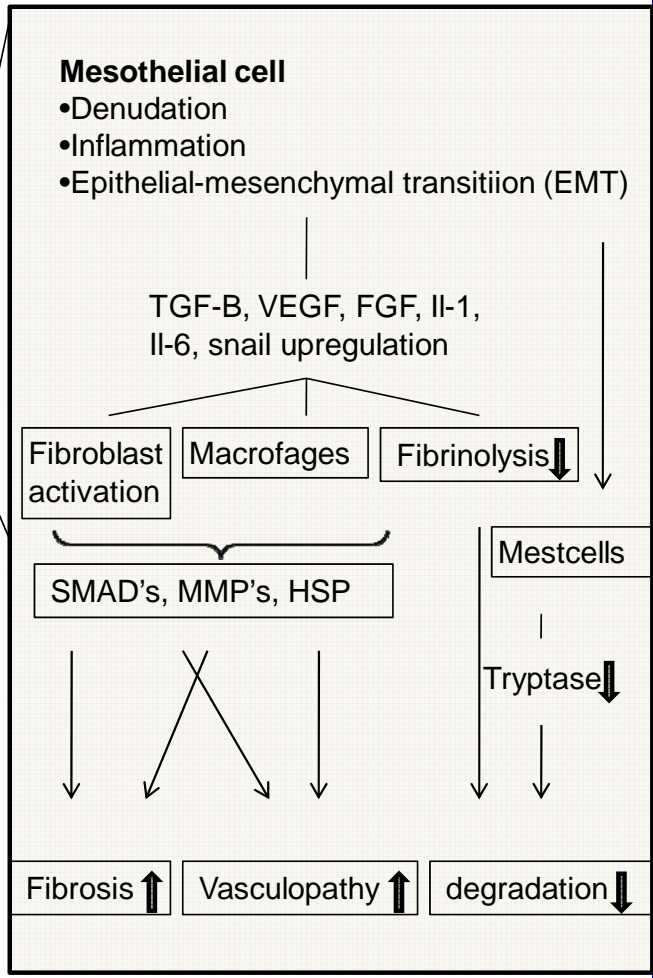
**Second hit – new riskfactors**

- PDF's: Glucose, GDP/AGE's, low pH, lactate buffer
- Transplantation
- Peritonitis
- Medication
- Unknown factor

peritoneum

Peritoneal remodelling

Simple sclerosis



Post-transplant EPS

“Classic” EPS



# Design

## Dutch initiative EPS registry

### Doctor friendly design

- Web based 6 monthly questionnaire to all Dutch nephrologists.
  - „ Did you have any suspicion of EPS in the previous 6 months? “
- If yes: short questionnaire is asked to fill in after consent

# Design

## Dutch initiative EPS registry

- EPS considered: research nurse will fill in CRF from patient records and interviews.
- Follow up: on site by research nurse twice a year.
- Compatible to other registries
- Additional collection serum, dialysate samples and material for marker studies and DNA research.

# Dataset variables EPS registry

## Diagnostic parameters

- Patient related: demographics, symptoms
- PD related
- Macroscopical and radiological investigations

## Prognostic parameters

- RRT modality, switches
- PD related: eg PET, peritonitis, fluids, markers
- Kidney transplantation related
- Nutrition status
- Chemistry

## Therapeutical parameters

- Medication, supportive and medication

Kidney TX	Comorbidity	Medication	Nutritional status	Blood chemistry	EPS treatment	Surgical procedures	
Basic details	Timeline	EPS symptoms	EPS investigations	PD treatment	PD adequacy	PET tests	Peritonitis

Baseline details

Study number

Centre

Date of inclusion

Date of Birth

Height

Sex  male  female

Ethnic Origin  Netherlands  other, specify

Country of birth  Netherlands  other, specify

Country of birth biological mother  Netherlands  other, specify

Country of birth biological father  Netherlands  other, specify

Ever smoked tobacco  yes  no  unknown

Current tobacco smoked  yes  no  unknown

Cause of renal failure  (EDTA Code)

Primary renal diagnosis  (EDTA Code)

Secondary diagnosis

Remarks

*Family disease history*

Diagnosis of renal disease in family  yes  no  unknown

Diagnosis of ischemic heart disease in family  yes  no  unknown



Study number 1  
Centre

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Kidney TX	Comorbidity	Medication	Nutritional status	Blood chemistry	EPS treatment	Surgical procedures	
Basic details	Timeline	EPS symptoms	EPS investigations	PD treatment	PD adequacy	PET tests	Peritonitis

Timeline

Number of RRT's

Remarks

Number	Start date	Modality	Reason switch	Reason switch, other specify
<input type="text" value="0"/>	<input type="text"/>	<input type="radio"/> HD <input type="radio"/> PD <input type="radio"/> TX	<input type="text"/>	<input type="text"/>



Study number 0

Centre 3

Find participant

Back to menu

Basic details

Timeline

EPS symptoms

EPS investigations

PD treatment

PD adequacy

PET tests

Peritonitis

Kidney TX

Comorbidity

Medication

Nutritional status

Blood chemistry

EPS treatment

Surgical procedures

### Surgical procedures

Any surgery that the patient has had involving the catheter, hernia's and abdominal surgery related or not related to PD. Special interest for parathyroidectomy.

Surgical procedures  yes  no

Remarks

Specify	Date	Outcome	Notes
<input type="checkbox"/> Catheter insertion (medical)	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Catheter insertion (surgical)	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Catheter reposition	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Catheter removal	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Laparotomy	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Omentectomy	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Parathyroidectomy	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Inguinal hernia repair	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Umbilical hernia repair	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Abdominal wall hernia repair	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Peritonolysis	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Adhesionolysis	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Bowel resection	<input type="text"/>	<input type="text"/>	<input type="text"/>
<input type="checkbox"/> Other, specify	<input type="text"/>	<input type="text"/>	<input type="text"/>



Study number 0

Centre 3

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Back to menu

Kidney TX	Comorbidity	Medication	Nutritional status	Blood chemistry	EPS treatment	Surgical procedures	
Basic details	Timeline	EPS symptoms	EPS investigations	PD treatment	PD adequacy	PET tests	Peritonitis

### Current clinical symptoms of EPS

Date EPS diagnosis  according to the physician

Duration of symptoms prior to diagnosis

Start date clinical symptoms  according to the physician

Start date clinical symptoms  according to the nurse

Nausea  yes  no  unknown

Appetite loss  yes  no  unknown

Weight loss  yes  no  unknown

Malnutrition  yes  no  unknown

Diarrhea  yes  no  unknown

Recurrent abdominal cramps  yes  no  unknown

Abdominal pain  yes  no  unknown

Vomitus  yes  no  unknown

Constipation  yes  no  unknown

Ileus/obstruction  yes  no  unknown

Ascites  yes  no  unknown

Remarks



Study number 0

Centre 3

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Back to menu

Kidney TX	Comorbidity	Medication	Nutritional status	Blood chemistry	EPS treatment	Surgical procedures	
Basic details	Timeline	EPS symptoms	EPS investigations	PD treatment	PD adequacy	PET tests	Peritonitis

EPS investigations

- Laparotomy peritoneum
- MRI-scan
- Investigations PD fluid: dialysates
- Other investigations
- X-rays
- Ultrasound scan
- Stored serum
- CT-scan
- Barium follow through
- Pathology: biopsies peritoneum

Remarks

Specify investigation

<u>Investigation</u>	<u>Date</u>	<u>Outcome</u>	<u>Reason</u>	<u>Remarks</u>
<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>

# Challenges EPS registry

- Uniform EPS diagnosis
- Different stages of EPS: determination what stages included
- Complete dataset with all possible variables
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[www.epsregistry.eu](http://www.epsregistry.eu)




EPS Registry - Windows Internet Explorer

http://www.epsregistry.eu/

Bestand Bewerken Beeld Favorieten Extra Help

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### EPS

Encapsulating peritoneal sclerosis (EPS) is a rare but serious complication of peritoneal dialysis (PD). This diagnosis needs to be considered in patients with a history of PD who are currently having or have experienced symptoms of recurrent partial or complete intestinal obstructions. The clinical assessment is difficult due to the insidious development of EPS. At this moment there is no clear test or radiologic investigation to detect EPS at an early stage. Risk factors for the development of the disease are incompletely understood.

The EPS registry is a collaboration of the Dutch Kidney centres and the Hans Mak Institute. Main aim of the nationwide registry is to improve the knowledge of the pathophysiology of EPS. Subsequently, uniform diagnostic criteria and management strategies will be developed. This registry has the potential to be extended to other European countries. There is already a close collaboration with the UK EPS registry.

The EPS registry is designed to register and follow patients with a possible and definite diagnosis of EPS. An inventory email questionnaire will be sent to all nephrologists in the Netherlands. We kindly ask all nephrologists to participate in this registry. When the diagnosis of EPS is considered, a research nurse will be deployed to the centre to gather all information on the patients. We encourage the treating physicians to report a patient even if there is only a slight suspicion of EPS. The information gathered will be reviewed by the steering committee of the registry. A yearly update, summarizing the data collected in the EPS registry, will be released on the website.

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### EPS case

If you considered the diagnosis of EPS in one or more patients with a history of PD, please complete this form. After submitting this form a research nurse from the Hans Mak Institute will contact you.

\* The asterisk fields are obliged to fill in.

Your name: \*

Hospital: \*

E-mail address: \*

Telephone number: \*

Question/remark:

Type the following code: \*

Submit

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# Started June 2009 First questionnaire

	Physicians	Dialysis Centres
Number of emails	259	60
Respos	113 (44%)	55 (92%)
EPS suspicion	27	19
EPS no	64	48
EPS colleague	23	12

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EPS

# Encapsulating Peritoneal Sclerosis patient

- Start PD at young age
- Long PD treatment (> 4 yrs)
- Insidious symptoms of intestinal tract:
  - Nausea, vomitus, weight and appetite loss
  - Inflammation
  - Abdominal pain and mass
  - Obstipation, ileus
- PD: fast transporter status, loss of ultrafiltration, bloody ascites
- Often after kidney transplantation
- 50% chance to die within 1 year.