به نام خدا


17th
International Congress
of Nephrology, Dialysis,
and Transplantation
Organized by Iranian Society of Nephrology
Tabriz , Iran 19-22 November 2019
CME Course
TABRIZ
2019
IPNA
MAYO
CLINIC
ISN

PODONET Consortium

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Isfahan University of Medical Sciences

With Permission of

Prof. Dr. Franz Schaefer

Prof. Dr. Dr. Franz Schaefer

Professor of Pediatrics

Head, Division of Pediatric Nephrology & KfH Children's Kidney Center.

Dr. Schaefer has founded and coordinates several international clinical research consortia such as the ESCAPE Clinical Research Network for Children with Chronic Kidney Disease, the International Pediatric Dialysis Network and the PodoNet Consortium for clinical, genetic and experimental podocyte research

Username

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PodoNet

Clinical, Genetic and Experimental Research into Hereditary Diseases of the Podocyte

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- Clinical Centers
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- PodoNet Registry Publications

EURenOmics

E-Rare

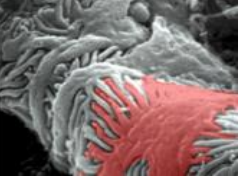
Welcome to PodoNet

The PodoNet Registry explores the demographics, causes and prognosis of patients with congenital and steroid resistant nephrotic syndrome. The Registry is open to all clinician scientists who want to contribute information about patients with this rare condition.

The clinical and genetic information collected serves to provide an evidence base for diagnostic and therapeutic decision-making, to establish genotype-phenotype correlations in hereditary forms of the disease, and to collect a critical mass of cases and families to foster the search for new genetic entities.

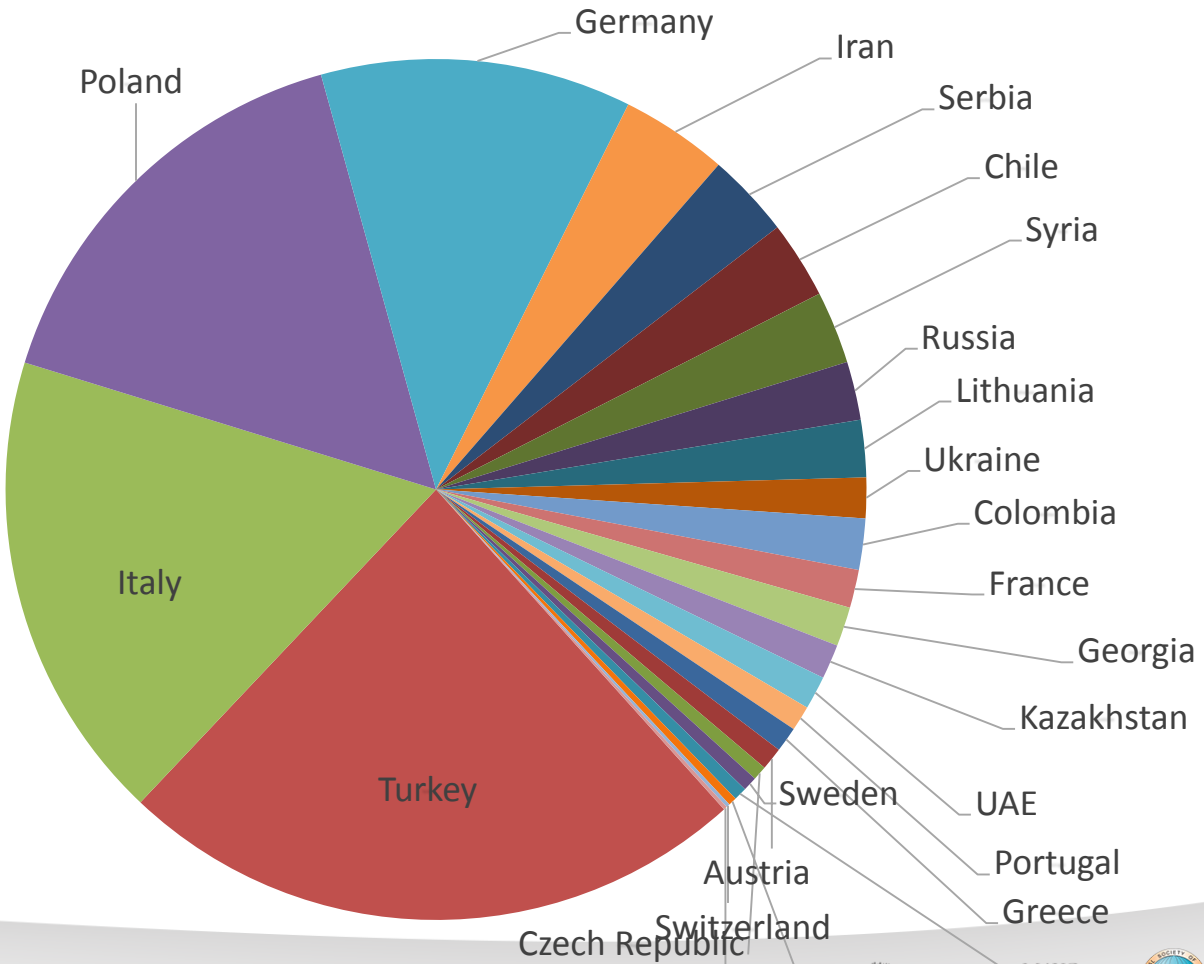
If you are a pediatric or adult nephrologist interested in participating in our registry, please complete this Registration Form.

To date, 101 pediatric nephrology centers in 31 countries have registered with PodoNet. Currently, 2254 patients from 80 centers are followed in the PodoNet registry.



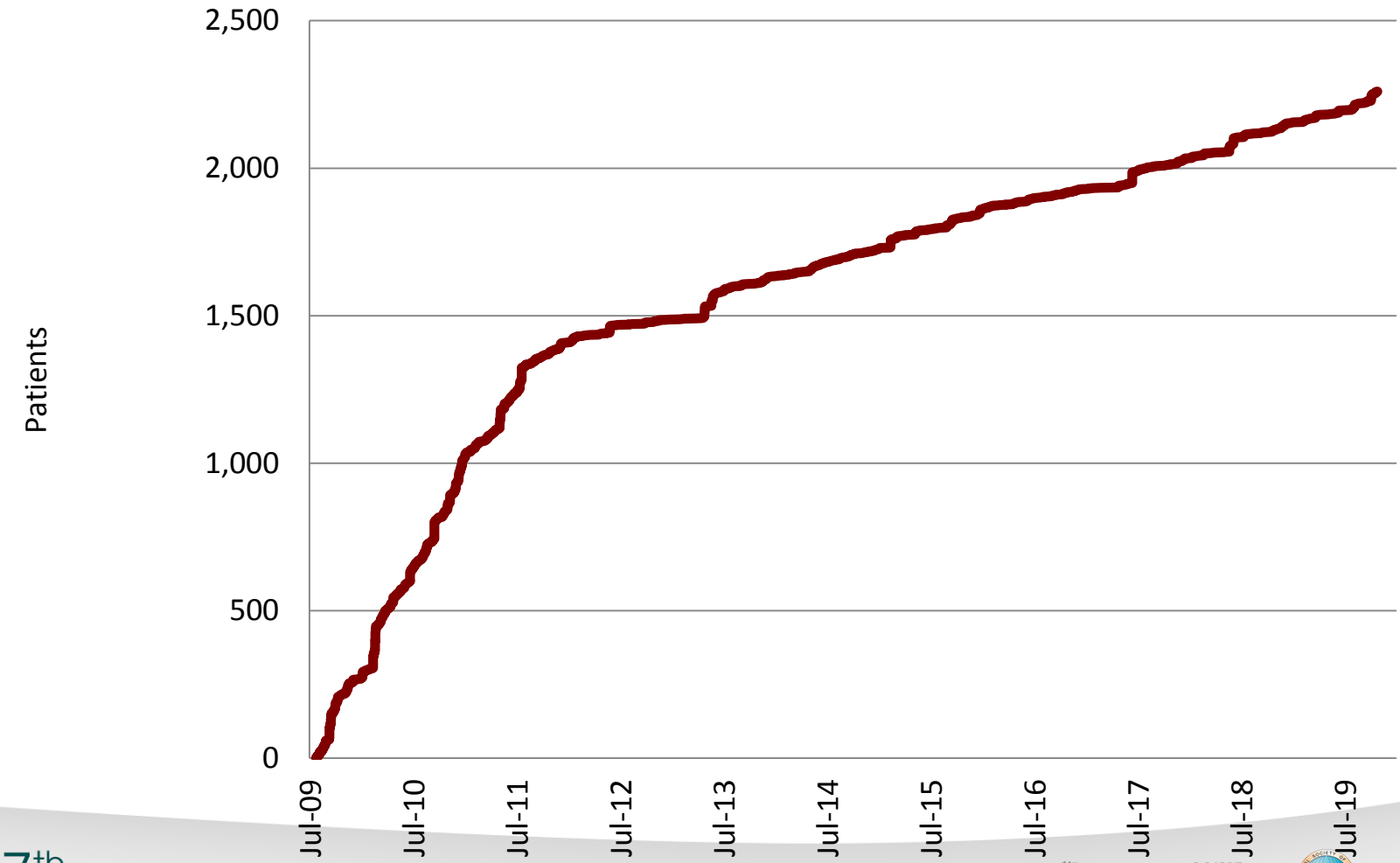
The PodoNet SRNS Registry

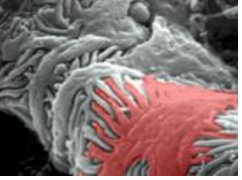
2205 children from 80 contributing centers in 28 countries



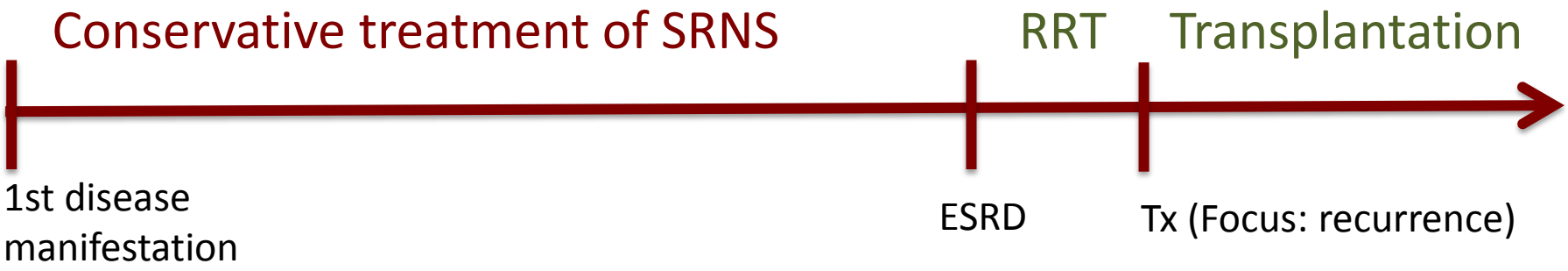
The PodoNet SRNS Registry

Patient Enrolment





Mixed Retro- and Prospective Data Collection

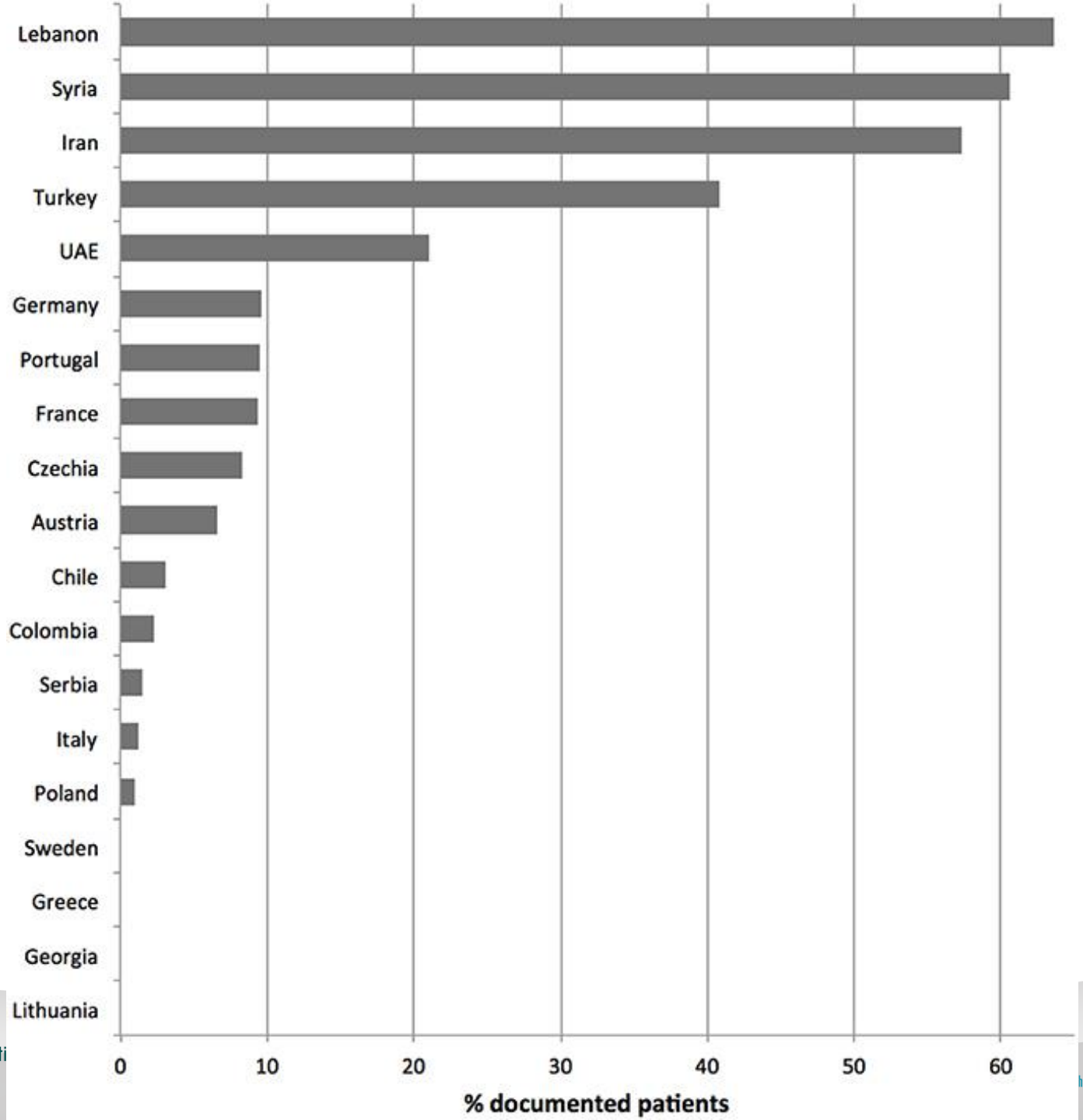


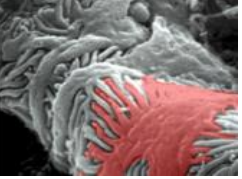
Basic characteristics:

- Info on 1st disease manif. 92%
- Kidney biopsy results 78%
- Genetic screening results 80% (NGS 45%, WES 3%)
- Family history 66%

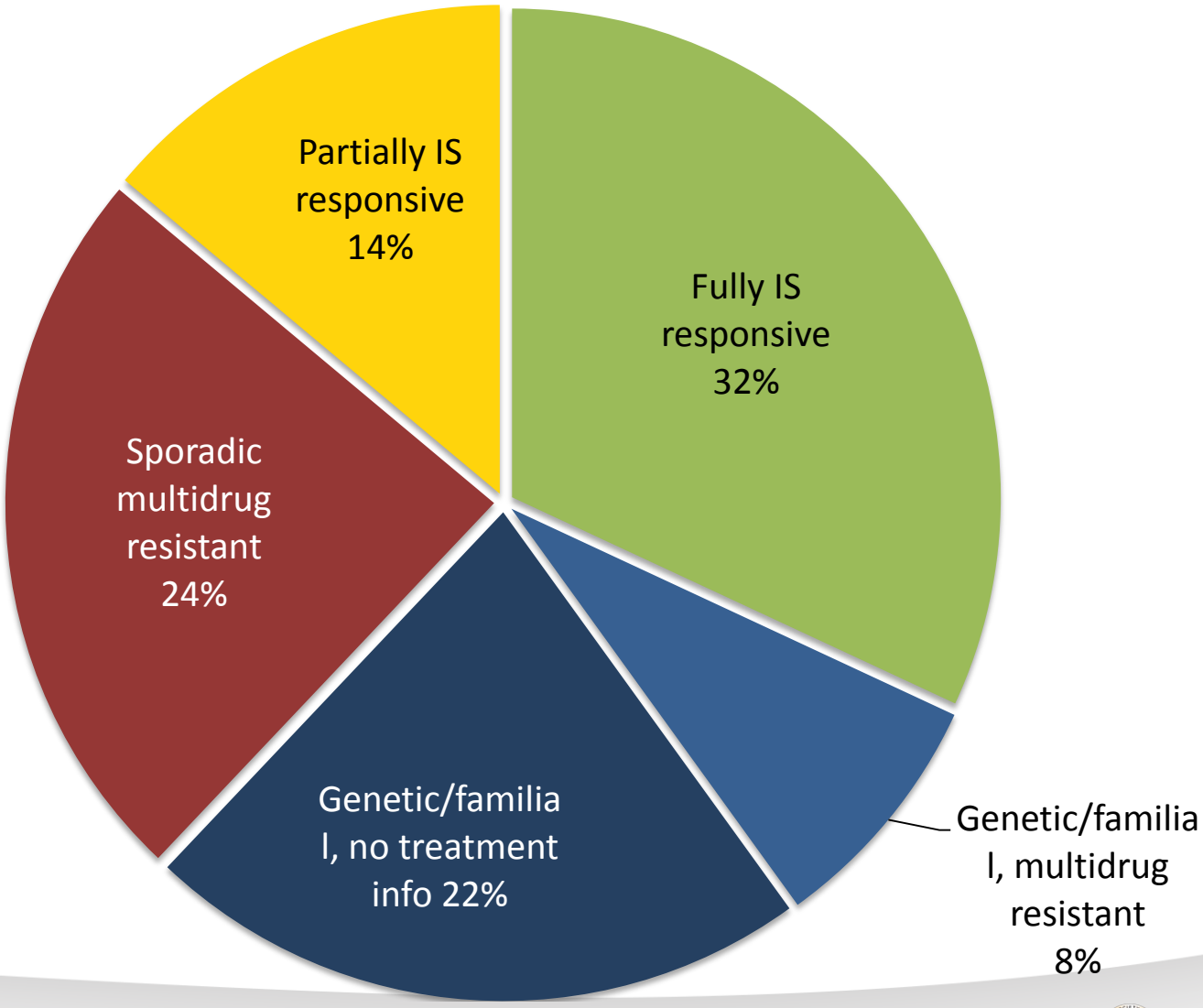
Follow-up:

- Clinical updates 7.7 ± 10.3/pt (N_{total}= 17.021)
- Medication updates 12.2 ± 19.4/pt (N_{total}= 26.891)
- Duration 5.6 (2.8; 10.0) years
- Progression to ESRD 641/2205 (29%) in 2.7 (0.9;5.6) years
- Post-Tx Recurrence 57/372 (15%)


Lebanon
Syria
Iran
Turkey
UAE
Germany
Portugal
France
Czechia
Austria
Chile
Colombia
Serbia
Italy
Poland
Sweden
Greece
Georgia
Lithuania
0
10
20
30
40
50
60
% documented patients

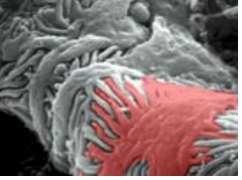


Distribution of SRNS Subtypes In **PodoNet** Cohort



N=1,168

Trautmann et al., Front Ped 2018



Pharmacotherapies Applied

MPR

CPH

MPR + CPH

CNI (CsA/ Tac) ± Pred ± RAS

RAS ± Pred

MPR

CPH

Ritu-ximab

CNI + MMF ± Pred ± RAS

MMF ± Pred ± RAS

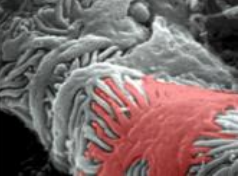
Multidrug-Resistant

IIS Responsive

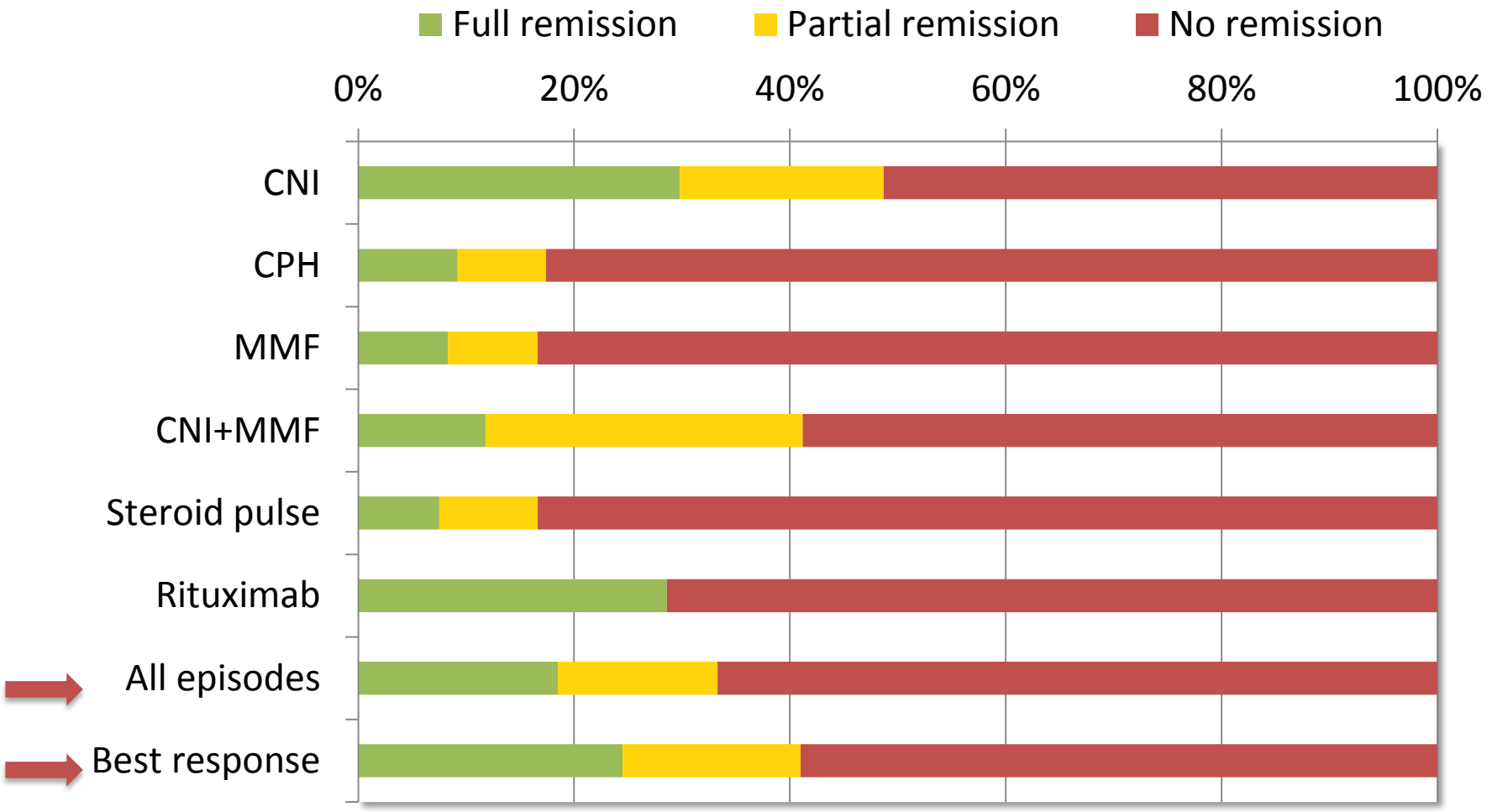
Responsive to IIS treatment

No responsiveness

Trautmann et al., Front Ped 2018

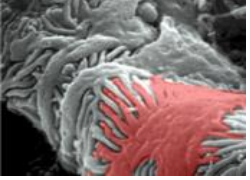


Treatment Response

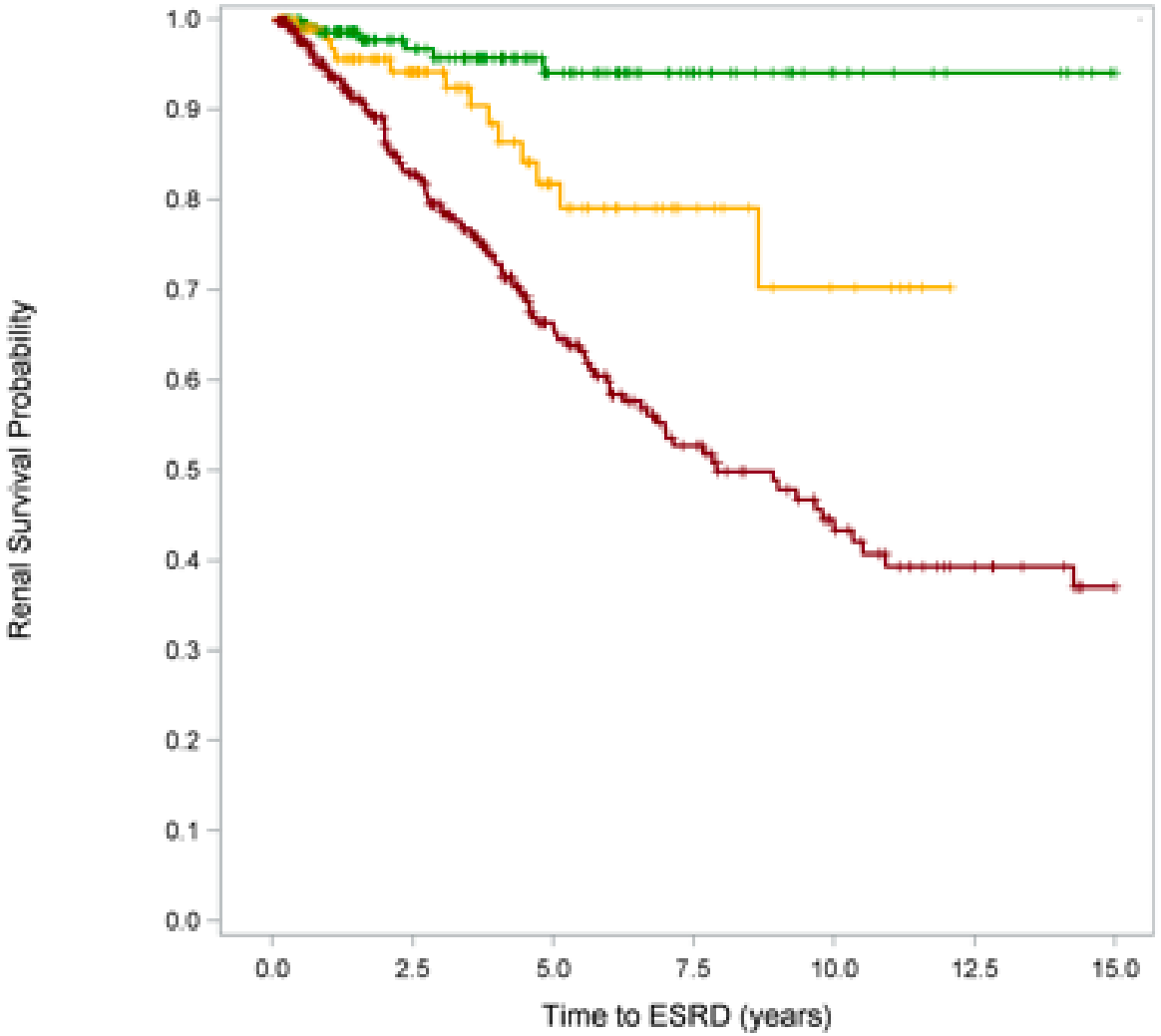


612 patients with 906 treatment episodes within first year after disease manifestation

Trautmann et al. JASN 2017



Response to Intensified Immunosuppression (IIS) Predicts Renal Prognosis



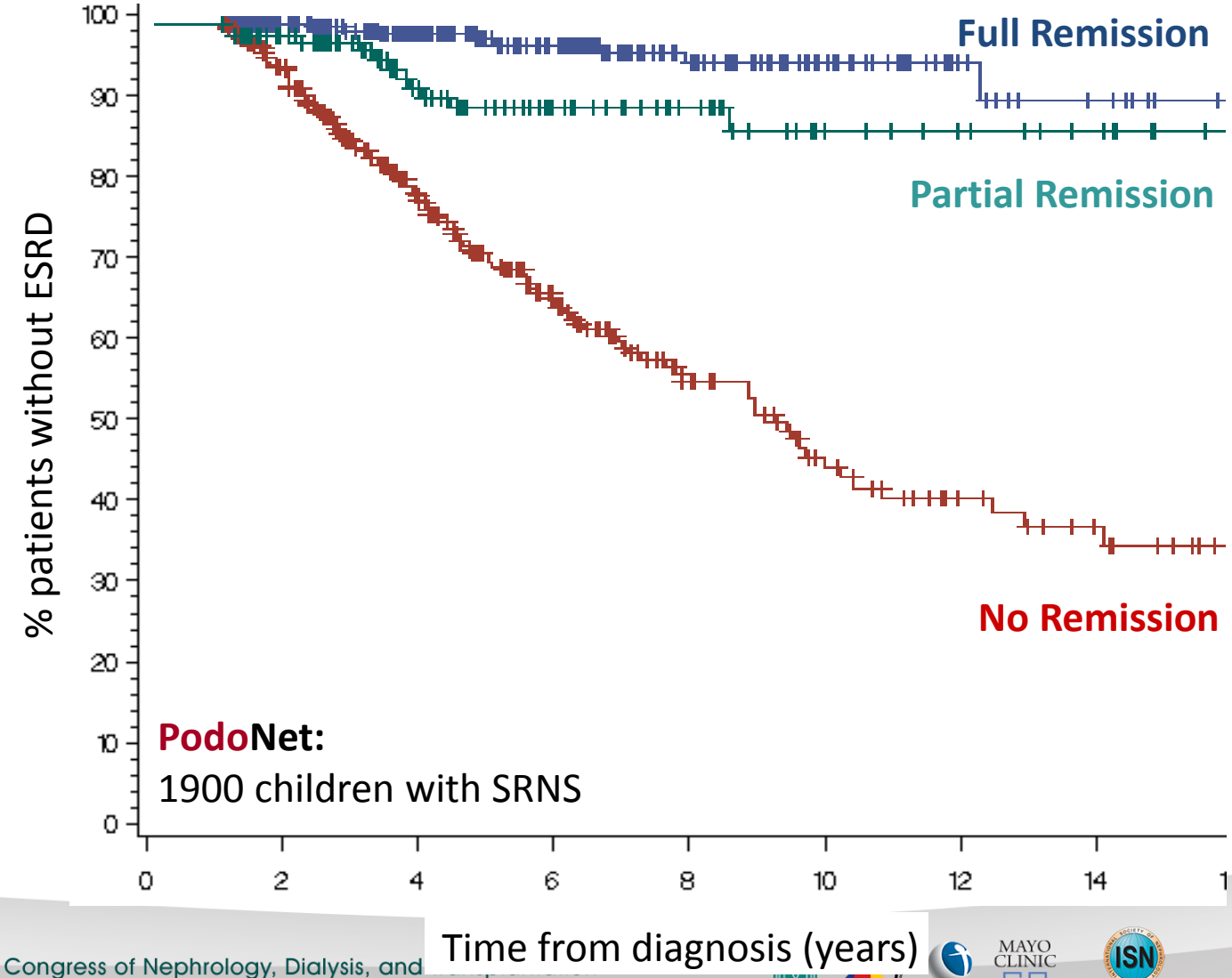
Full Remission

Partial Remission

No Remission

Trautmann et al. JASN 2017

Response to Intensified Immunosuppression Predicts Renal Prognosis



Trautmann et al. JASN 2017

NGS Podocytopathy Gene Panel Screening

Known Genes(n=30)

AD genes: *ACTN4, ANLN, INF2, TRPC6, ARHGAP24*

AR genes: *NPHS1, NPHS2, PLCE1, MYO1E, DGKE, CD2AP, PTPRO, CRB2, EMP2*

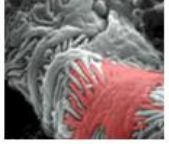
Syndromic genes: *LAMB2, LMX1B, GLA, SMARCAL1, WT1, ITGA3, PAX2, WDR73*

Collagenopathy genes: *COL4A3 COL4A4 COL4A5*

Mitochodropathies: *COQ2, COQ6, PDSS2, MT-TL1, ADCK4*

Candidate Genes (n=11)

APOL1, C14orf142, ITGB4, KANK2, MAGI2, MPDZ, MYH9, CD151, TTC21B, SCARB2, ARHGDIA



PodoNet

Clinical, Genetic and Experimental Research into Hereditary Diseases of the Podocyte

Genetic diagnosis made in 333 of 1,294 SRNS subjects (25.7%)

Mutation detection rate:

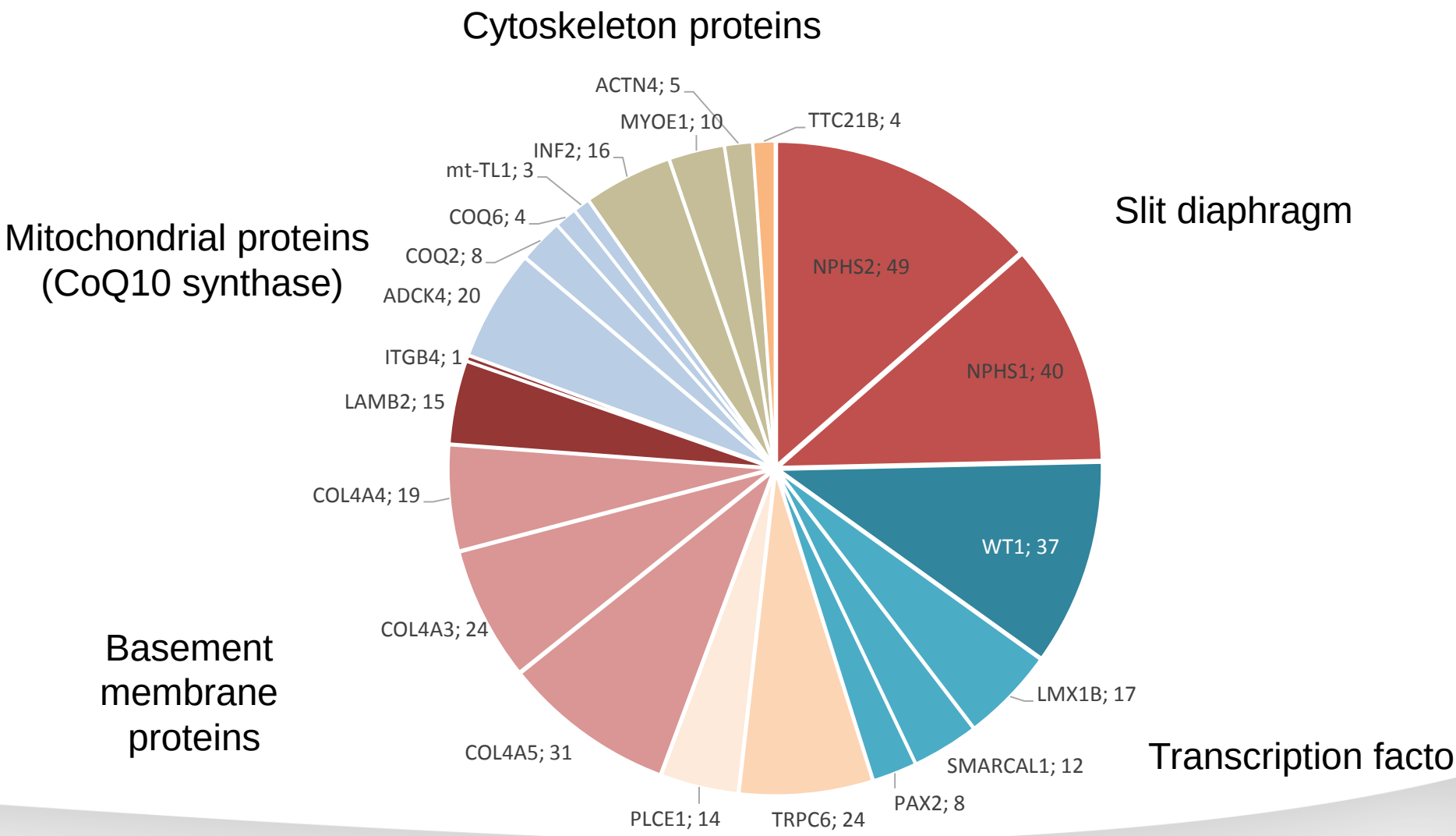
41% of familial cases

36% of sporadic but consanguineous cases

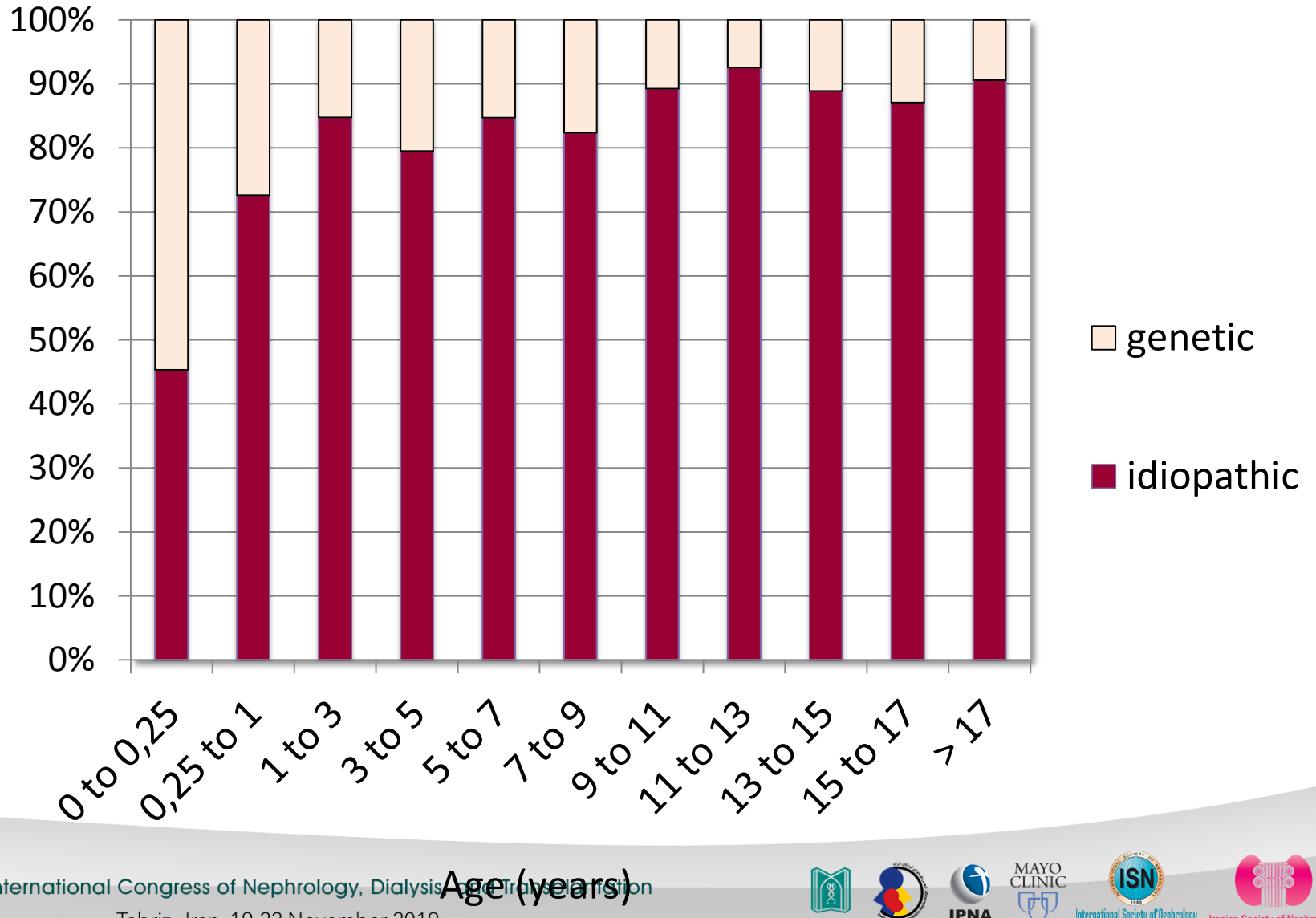
19% of sporadic, non-consanguineous cases

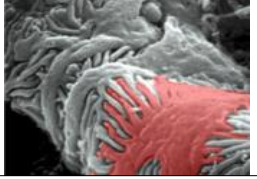
Gene	Classification
NPHS1	1st line SRNS gene
NPHS2	1st line SRNS gene
WT1	1st line SRNS gene ; syndromic gene
INF2	well-acknowledged causative gene
PLCE1	well-acknowledged causative gene
TRPC6	well-acknowledged causative gene
CD2AP	well-acknowledged causative gene
ACTN4	well-acknowledged causative gene
COQ2	syndromic gene
COQ6	syndromic gene
LAMB2	syndromic gene
LMX1B	syndromic gene
SMARCAL1	syndromic gene
ADCK4	novel gene proposed in a few recent studies
ARHGDIA	novel gene proposed in a few recent studies
ITGA3	novel gene proposed in a few recent studies
MYO1E	novel gene proposed in a few recent studies
MYH9	novel gene proposed in a few recent studies
PTPRO	novel gene proposed in a few recent studies
C14ORF142	novel gene proposed in a few recent studies
CD151	novel gene proposed in a few recent studies
EMP2	novel gene proposed in a few recent studies
PDSS2	novel gene proposed in a few recent studies
SCARB2	novel gene proposed in a few recent studies
ARHGAP24	candidate gene
KANK2	candidate gene
ITGB4	candidate gene
MAGI2	candidate gene
MPDZ	candidate gene
TTC21B	candidate gene
mt-TL1	candidate gene

Podocytopathy Genes

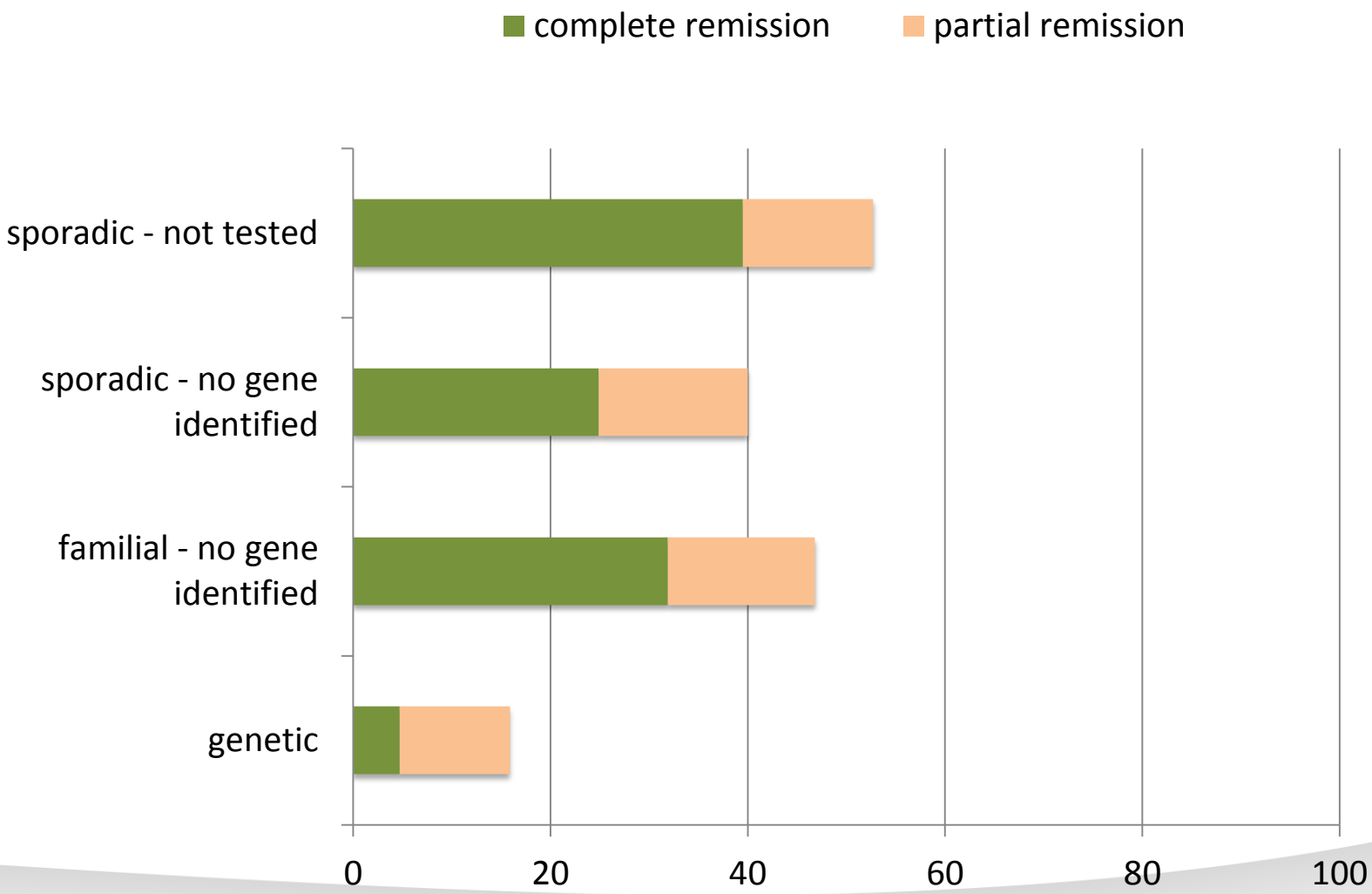


Genetic Forms of SRNS: Age at Manifestation

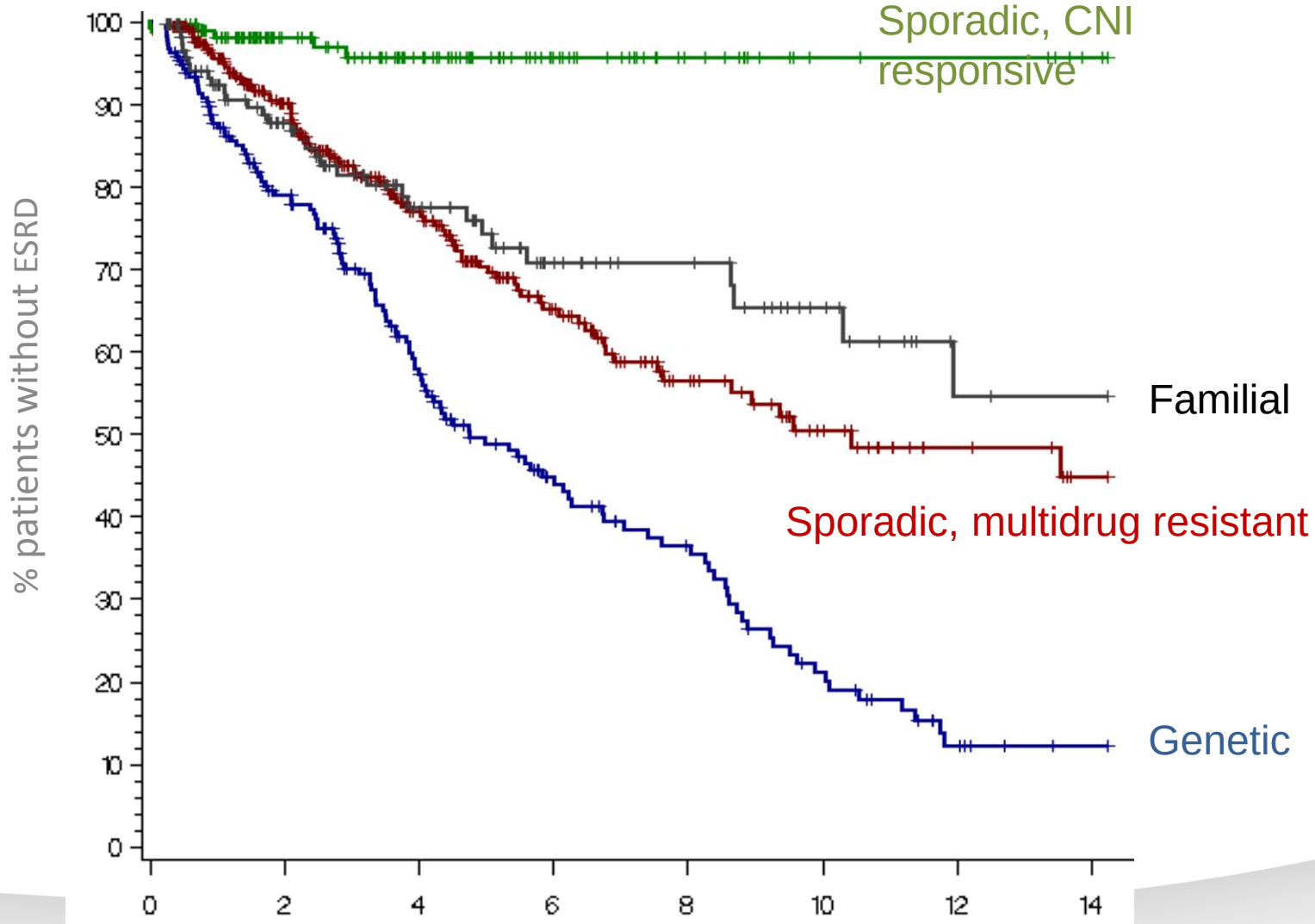




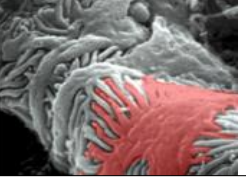
Response to Intensified Immunosuppression by Genetic Status



Prognostic Impact of Genetic Diagnosis



Trautmann et al. JASN 2017



Genetic Findings Predict Post-Tx Recurrence Risk

1766 patients
with primary SRNS

260 transplant recipients

233 with recurrence information

Recurrence	Yes n=37	No n=196
idiopathic	29	76
genetic	4	103
unknown	4	17

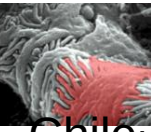
Idiopathic
n=105
(49.5%)

Recurrence rate
27.6%

Recurrence rate
15.8 %

Genetic
n=107
(50.5%)

Recurrence rate
3.7 %



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THANK YOU :)