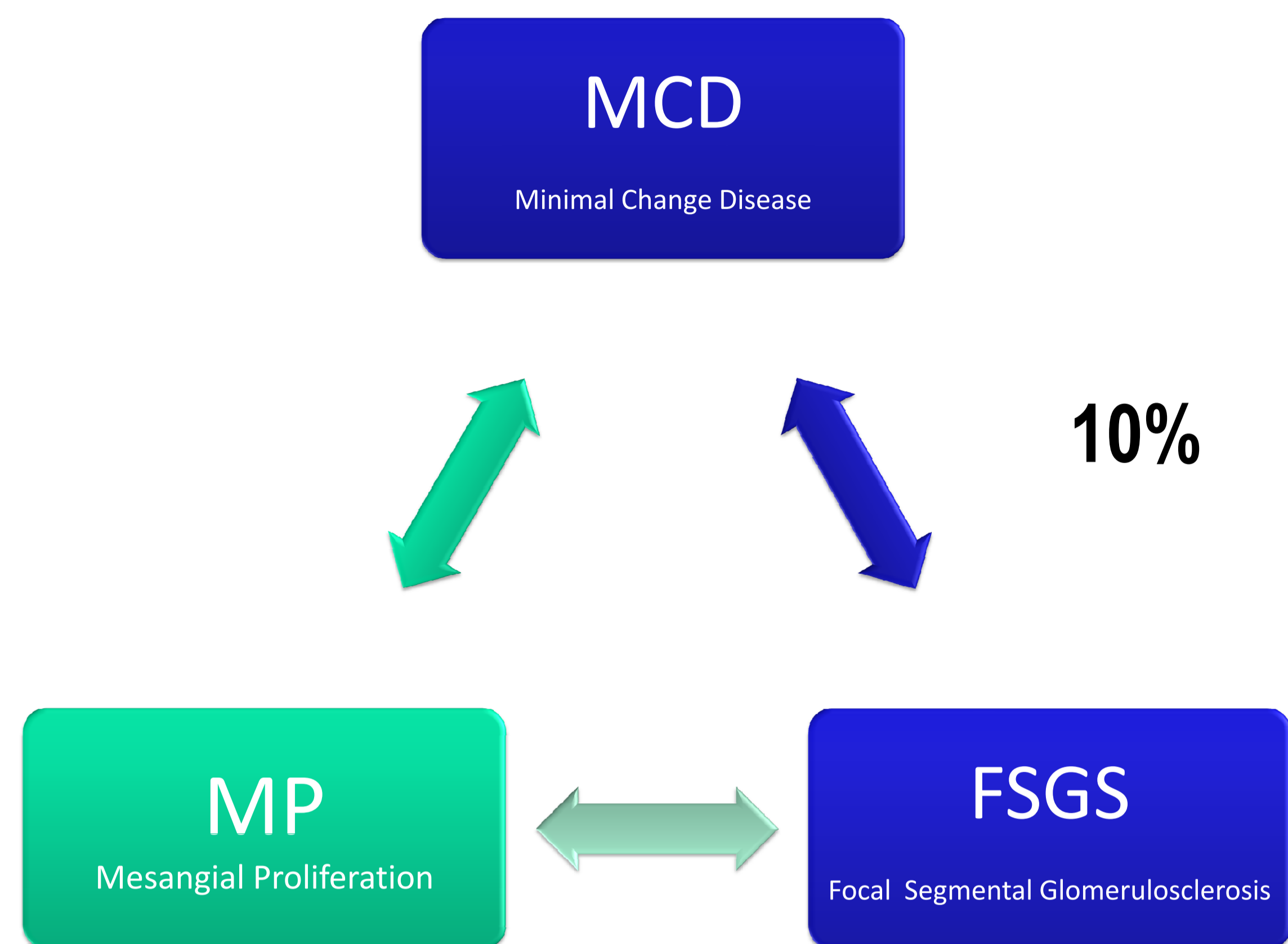


Natural history of Nephrotic Syndrome in Infancy



Introduction

FSGS is the most frequent acquired entity that leads to end stage renal failure in children (10 – 12%). The risk of recurrence post Tx is 20 – 50%. The risk of graft loss is 30 – 50%.

PREVIOUS REPORTS USING PL AND HIGH DOSES OF CYCLOSPORINE

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PL pre Tx using high dose CyA is proposed as an effective protocol in diminishing recurrence and increasing remission of recurrence when it appears immediate post Tx. We report our experience based in the Children's Hospital Boston CHB protocol, which combines PL, high dose CyA (IV and Oral) and Cyclophosphamide CP during the 2 months following Tx.

Methods

We reviewed records from 16 pts who went to 17 KTx with primary FSGS (1998-2012). In living donors-LD- we used PL #3 and CyA IV previous to Tx. Recurrence was defined as a serum albumin level < 3.0 grs/lit and a urine Prot/creat ratio > 1. The treatment: PL every day #4, then every other day up to 15 sessions , CyA at high dose (8-10 mg/kg/day) to maintain CyA serum levels C2 on the highest range allowed, CP 1-2 mgrs/kg/day for 2 -3 months and regular steroids. All of them received induction therapy with Daclizumab Basiliximab or Thymoglobulin , and 6 months prophylaxis for CMV.

Total remission was defined as urine Prot/creat ratio < 0.5 and serum albumin level > 3 grs/lit.

Carachteristics	Values
Age at diagnosis, median (IQR) years	6.25 (5.5 – 9)
Age at Transplant, mean (range) years	13 (4.7 – 19)
Interval: diagnosis - transplant, median (IQR) years	7.23 (3.8 – 8.9)
Gender, n male	10/16
Previous nephrectomy, n	10/17
Type of dialysis	
Hemodialysis, n	9/17
DP	8/17
Type of Donor , n	
Cadaveric	8/17
Live	9/17

Carachteristics	(n=17)
Induction, n	
Thymo	6
Basiliximab	10
Daclizumab	1
Immunosuppression, n	
Cyclosporine	14
Cyclophosphamide	13
Steroids	17
Tacrolimus*	1
MMF*	4
Rituximab	2

Results

Carachteristics	Values
Recurrence, n	15/17
Remission, n	11/15
Plasmapheresis, n	15
Mean (range)	13 (4.7 – 19)
ATN, n	6/17
AR , n	2/17
Infections, n	6/17
Prot/Creat urine ratio, median (IQR)	
Initial	7.9 (4.5-11.8)
At clinic discharge	0.3 (0.25-0.6)
3-months after	0.3 (0.25-0.6)
Creatinine 3 months after, median (IQR)	1.1 (0.82-1.8)
Hospital stay , median (IQR) days	22 (16 – 39)

LD recurrence	8/9	88.8%
LD remission	7/8	87.5%
CD recurrence	7/8	87.5%
CD remission	5/8	67.5% went to remission
	2/8	25% died
	1/8	12.5% failed
Recurrence	13/17	88.8%
Total remission	11/15	73.3%

Conclusions

- FSGS´s recurrence post Tx was 15/17 (88.8%) . Response to treatment based in PL and high dose CyA is effective 11/15 (73.3%) .
- Level of urine Prot/creat ratio seems to be less in the recurrence when using pre KTx PL and CyA i.v.
- Pts were less likely to do ATN. It needs more # patients to corroborate it .
- Nephrotic Syndrome FSGS has a high recurrence in our population.
- Genetic studies were not practiced.

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