2nd International Conference

HUS-MPGN-PNH

Current diagnosis and therapy of thrombotic microangiopathies: hemolytic uremic syndrome (HUS), membrano proliferative Glomerulonephritis (MPGN) and paroxysmal nocturnal Hemoglobinuria (PNH)

Abstract form (in English)

Title (in capitals)

Authors//Institution/ Department

T e x t Structure:
The aim of the study
Methods
Results
Conclusion
Please, type using the **Times New Roman, large 12**

To be sent to:
Prof. LB. Zimmerhackl, Dptm. of Paediatrics Anichstraße 35. A-6020 Innsbruck, using the E-mail and paralelly the air/surface mail, **dead-line May 3, 2010!**

SUCCESSFUL KIDNEY TRANSPLANTATION IN 4 PATIENTS WITH FACTOR H DEFICIENCY-HUS

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Aim of the study: Factor H deficiency hemolitic uremic syndrome (FHD-HUS) has a very high risk of recurrence after kidney transplantation (KTx). Refraining from KTx, combined liver-KTx or KTx followed by lifelong plasmaexchange (PLE) have been proposed for patients with ESRD due to FHD-HUS with contrasting results. Herein we describe our protocol for KTx in FHD-HUS, which proved to be successful in all the patients we have treated so far.

Methods: Four patients (age range 5-36 yrs) with CKD5 due to documented FHD-HUS underwent KTx following one PLE before KTx and several maintenance PLEs and fresh frozen plasma infusions after KTx, according to the protocol shown in the table:

Time	PLE	Plasma Infusion	Frequency
Pre-KTx	75ml/kg	1000 ml	Once
POD 1-5	75ml/kg	no	Daily
POD 6-7	50ml/kg	no	Daily
POD 8-17	50ml/kg	25ml/kg	Alternate day
POD 18 to 26	50ml/kg	no	Every other day
POD 27-41	50ml/kg	no	Every 5 days
POD 42 to 180	STOP	20ml/kg	Weekly

Legend: POD: Post KTx day

Immunosuppressive protocol included basiliximab, tacrolimus or cyclosporine, MMF and prednisone. We emphatize that all patients were addressed to KTx with a significant fluid overload (as much as 3% above optimal body weight) obtained with plasma infusion.

Results: Over a cumulative observation period of 61 mos., we observed only 2 recurrences (in 2 different patients) which were managed with PLE (1 case) and Eculizumab (1 case) with immediate recovery from the recurrence.

Conclusions: Our therapeutic approach to TKx in FHD-HUS represents a less aggressive solution in the meantime that Factor H becomes available for maintenance treatment.

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