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!**

HEMOCONCENTRATION IS MAJOR RISK FACTOR FOR CENTRAL NERVOUS SYSTEM INVOLVEMENT IN HEMOLYTIC UREMIC SYNDROME

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Aim of the study: HUS, in its typical variant (D+), is an unrare thrombotic microangiopathy (TMA) and severe central nervous system involvement (CNSI) accounts for the vast majority of the mortality and part of the long-term most severe sequelae.

Methods: The charts of all D+HUS cases referred to our network during the last 10 years were reviewed, to test the working hypothesis that CNSI is associated with more severe hemoconcentration (HC) at disease onset. A total of 61 D+ HUS cases (27 males) were traced and retrospectively grouped as follows: I)Patients (Pts) who did not require renal replacement therapy (RRT) w/o CNSI; II)Pts who required RRT w/o CNSI; III)Pts with CNSI. The groups were compared (ANOVA) for general demography and basic biochemestry at admission.

CHARACTERISTICS OF PATIENTS AT PRESENTATION ACCORDING TO OUTCOME

	I	II	III	p
Age (Yrs)	3.0±3.0	3.8±3.2	3.7±3.0	0.61
HB (g/dL)	7.5 ±1.9	9.4 ±2.1	11.2 ±2.3	<0.0001
PTL (x10 /mm3)	81.9 ±73.4	87.8 ±67.9	79.9 ±59.4	0.93
sCr (mg/dL)	1.33 ±1.03	4.24 ±2.55	4.05 ±3.06	<0.0001

Results: Results show a surprising direct correlation between Hb level and disease severity: pts with CNSI (Group III) had a significantly higher Hb level compared to both group I and II.

Conclusions: In the presence of acute hemolysis a less severe anemia should be interpreted as sign of HC which, in combination with TMA, may lead to more severe tissue hypoperfusion and ischemic organ damage. In conclusion, high Hb level seems a major risk factor for subsequent CNSI and, whenever detected, dehydration should be actively ruled out and corrected; fluid restriction in these patients may be very dangerous.

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