

## **D+ hemolytic-uremic syndrome: A systematic review**

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### **Abstract**

Hemolytic-uremic syndrome (HUS) is the most common cause of acute renal failure in children below 3 years of age. It is defined by a triad of symptoms which associates hemolytic anemia with fragmented erythrocytes, thrombocytopenia and acute renal failure. Three types of HUS can be distinguished: typical HUS also called diarrhoea-associated (D+) HUS, atypical HUS (a-HUS/ D-HUS) and secondary HUS (drug induced, C+HUS, in patients receiving marrow transplantation, post partum etc.). The common event among these entities appears to be vascular endothelial cell injury, which induces mechanical destruction of erythrocytes, activation of platelet aggregation and local intravascular coagulation, especially in the renal microvasculature. D+ HUS represents nearly 90% of HUS in children. Evidence of exposure to Shiga toxin (STx 1, 2) producing *Escherichia coli* (STEC) - also called Enterohemorrhagic *Escherichia coli* (EHEC) - has been demonstrated in many countries in about 85% of cases. Recently serotypes O157:H7, O26 are the most frequent. Early and accurate supportive treatment and early start of dialysis is the major importance and allows a current mortality rate below 5%. Vital prognosis is compromised in cases with CNS or multi-visceral involvement. After 15 years or more of apparent recovery, 20% to 60% of patients have residual renal symptoms, with up to 20% having chronic renal insufficiency (CRI) or end-stage renal disease (ESRD). Atypical HUS represents less than 10% of HUS in children. Some of these cases (familial) are associated with low C3 levels, hereditary deficiency of factor H or with mutations in factor H gene. The deficiency of von Willebrand factor cleaving protease deficiency in D+HUS, as reported in adults with thrombotic thrombocytopenic purpura (TTP), is not present.

### **Biography**

Květa Bláhová is an Associate Professor in the Department of Pediatrics at Charles University and University Hospital Motol in Prague. She received many awards. She has membership in prestigious European societies. She has published numerous papers in reputed journals.



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Nephrologists-2016

# Nephrologists Annual Meeting

March 31-April 02, 2016 Valencia, Spain