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Saudi children have high prevalence of genetic related atypical hemolytic uremic syndrome and better renal recovery with eculizumab therapy

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Background & Aim: Atypical hemolytic uremic syndrome (aHUS) is ultra-rare disease, characterized by microangiopathy hemolytic anemia, thrombocytopenia and renal impairment. Genetic defects that determine uncontrolled activation of the alternative complement pathway have been well documented, it accounts for about 40%–60% of the cases. In Saudi Arabia, there is up to 56% of consanguinity marriage resulting in higher prevalence of genetic diseases. We are reporting the experience of a tertiary care center in Saudi for children with aHUS who were treated their outcome and genetic background.

Methods: This is a retrospective study, from 2010 till May 2017 in a tertiary care center comparing children with aHUS who had received plasma therapy to those received Eculizumab therapy which was introduced at our center in 2014.

Results: 21 Saudi children who have similar demographic background diagnosed with aHUS, 12 (57%) of them showed complete renal and hematologic recovery (67% in the Eculizumab group versus 33% in plasma therapy group). Six cases (29%) reached End Stage Renal Disease (ESRD), four patients (67%) of these cases from the plasma therapy group; two patients (33%) from Eculizumab group reached ESRD, their genetic mutations were not related to complement dysregulation system. Two of the 21 cases (17%) developed disease recurrence while receiving plasma therapy but no recurrence developed after using eculizumab. 11 (69%) of the 16 cases who underwent genetic testing has identified gene mutations.

Conclusion: In our 21 cases with aHUS, eculizumab was superior to plasma therapy in inducing, maintaining remission, and associated with better renal recovery. Genetic mutations detected among our patients were higher than reported for this ultra-rare disease, most probably related to the high prevalence of consanguinity marriage.

Biography

Abdulaziz Ahmed Bamhrazhas completed his MBBS from Hadramout University of Science and Technology and have Saudi & Arab Board of General Pediatrics. I am doing my pediatric nephrology fellowship programm under Saudi council for health specialties at king fahad medical city -Riyadh.

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