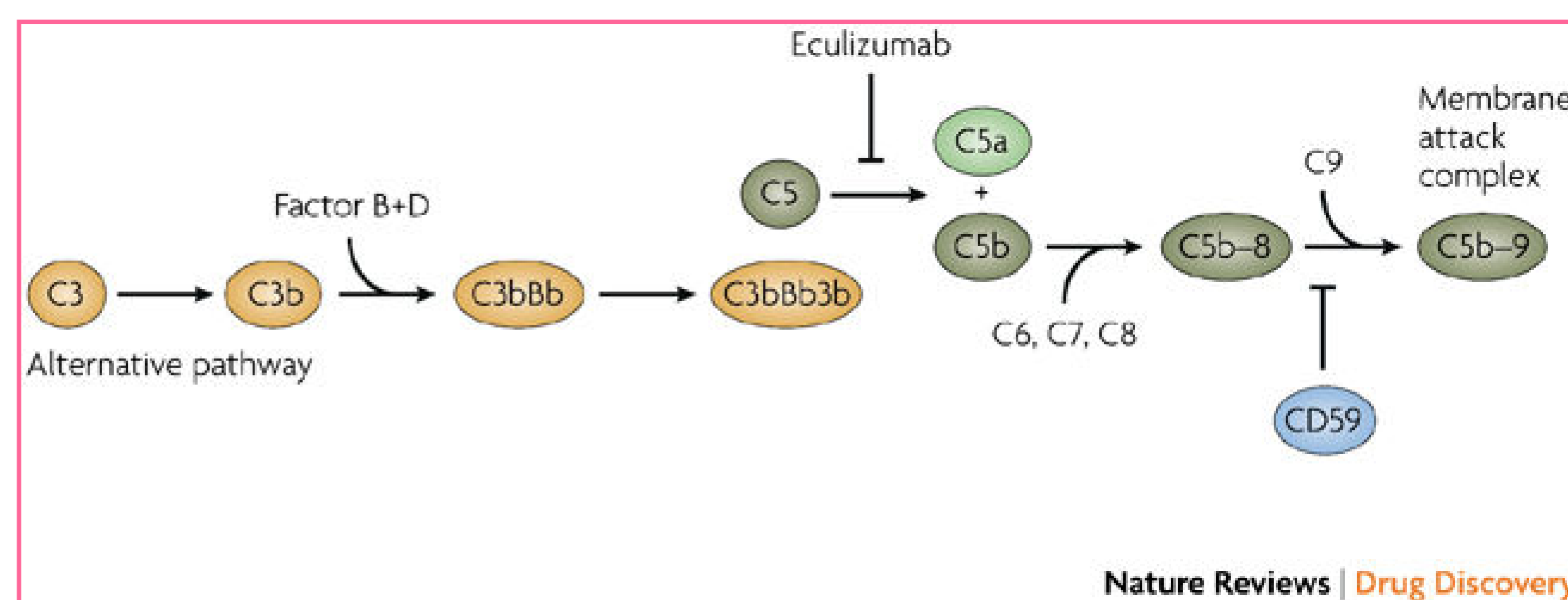


4CPS-163 ECULIZUMAB THERAPY FOR ADULT RENAL TRANSPLANT IN aHUS WITH MUTATION IN THE CFH GENE: A CASE REPORT

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BACKGROUND

Haemolytic uraemic syndrome (HUS) is a clinical entity defined as the triad of nonimmune haemolytic anaemia, thrombocytopenia, and acute renal failure, in which the underlying lesions are mediated by systemic thrombotic microangiopathy (TMA). Atypical HUS (aHUS) is a consequence of the insufficient regulation of the activation of the complement on cell surfaces, leading to endothelial damage mediated by C5 and the complement terminal pathway. Eculizumab is a monoclonal antibody that inhibits the activation of C5 and blocks the formation of the cell membrane attack complex. It has been associated with significant long-term improvements in renal function and important reductions in the need of dialysis.



PURPOSE

To describe a case of aHUS in which eculizumab is used as prophylactic therapy to prevent post-transplant aHUS recurrence.

MATERIAL AND METHODS

Retrospective case report of a 36-years-old female diagnosed with aHUS who carried the heterozygous mutation c.2557T>C in the CFH (Complement Factor H) gene.

RESULTS

	2010	2016	2017
Diagnosis	Atypical haemolytic uraemic syndrome.	Atypical haemolytic uraemic syndrome	Atypical haemolytic uraemic syndrome
Symptoms	Haemolytic anaemia, acute renal failure, hypertension, cardiorrespiratory arrest.	Chronic kidney disease.	Chronic kidney disease.
Initial treatments	Losartan 100 mg/daily, plasmapheresis sessions, prednisone, dialysis, two doses of eculizumab.		Kidney transplantation. Induction schedule: thymoglobulin, prednisone, mycophenolate-mofetil, tacrolimus, prophylactic eculizumab (first dose 1200 mg 6h pre-transplant, then within 24h and then three weekly doses of 900 mg).
Maintenance treatments	Dialysis, calcium acetate, cholecalciferol 0.266 mg/bimonthly, losartan 100 mg/daily.	Dialysis, losartan 25 mg/daily, paricalcitol 1 mcg/daily, darbepoetin alpha 20 mcg/weekly, iron sucrose 50 mg/weekly.	Eculizumab 900 mg in fortnightly cycles. Immunosuppressive therapy: prednisone, tacrolimus, mycophenolate-mofetil. Darbepoetin alpha 40 mcg/weekly. Iron sulphate 100 mg/daily. Prophylactic penicilin, valganciclovir 450 mg/daily.

After sixteen weeks, this patient has an adequate renal function (creatinine 0.78-mg/dl and glomerular filtration 90-ml/min/1.73m²).

CONCLUSION

This case adds to the evidence of the efficacy of eculizumab prior to kidney transplantation in preventing progression of aHUS post-transplant and without the need for plasmapheresis.

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L04 - Immunosuppressive agents