Bloody diarrhoea is a result of haemorrhagic colitis. AKI develops over 1 week as a result of injury of the renal vascular endothelium secondary to toxin production.

Endothelium damage leads to excessive platelet aggregation, platelet microvascular thrombi and AKI

Epidemic form associated with a prodromal illness and bloody diarrhoea. Follows infection with verotoxin producing enterococci (E.coli 0157) or shigella

Atypical HUS is rarer and has a poorer prognosis. It may occur following:
1 - Strep. pneumoniae, CMV or HIV infection
2 - Bone marrow or solid organ transplant
3 - Drug exposure (quinine, heroin, ciclosporin)
4 - Malignancy
5 - Pregnancy

Investigations:
- FBC and film - reticulocytes, haemolysis and thrombocytopaenia
- Direct Coombs test (differentiate between immune and non-immune)
- LDH (raised in haemolysis)
- U+E
- LFTs (including split bilirubin)
- Clotting
- HIV and hepatitis serology
- Autoimmune and vasculitis screen
- Stool MC+S
- Urinalysis
- Renal imaging

Mortality for HUS is 3-5%. 70-85% of epidemic HUS recover normal renal function.
Atypical HUS has a poorer prognosis initial mortality 25% and up to 50% progressing to ESRD

A triad of:
1 - MAHA
2 - Thrombocytopaenia
3- Renal failure

Haemolytic uraemic syndrome
Thrombotic thrombocytopaenic purpura

A pentad of:
- Thrombocytopaenia
- MAHA
- Fluctuating neurological signs (due to endothelial damage in cerebral circulation
- Renal Impairment
- Fever

Management
- Administer oxygen as needed
- Careful fluid therapy and electrolyte balance
- CVS support
- Renal and haematology input
- Treat the cause
- Plasma exchange - often used as difficult to distinguish from TTP
- Recommended in atypical HUS
- In epidemic HUS, PEx, IVIg, steroids and antiplatelets have not proven benefit

Plasma exchange:
Using FFP deficient in ultra large vWF
Should continue for at least 2 days after platelet recovery.
Removes the autoantibodies and replaces vWF-CP

Rituximab - Monoclonal antibody against CD20

Adjuvant high dose pulsed methylpred - No RCT evidence.

Platelet transfusion are contra-indicated unless there is life threatening haemorrhage as it worsens thrombosis

Genetic or acquired absence of von Willebrand factor cleaving protease, or ADAMTS13

vWF is a large glycoprotein in the plasma. binds factor VIII and activating and binding platelets in endothelial injury.
Produced in the endothelium as large multimers and inactivated when cleaved by vWF-CP.
In TTP multimers aren't cleaved leading to uncontrolled platelet activation, fibrin deposition and thrombus propogation

Low dose aspirin when platelets >50