

Table W2 Treatment and outcome of patients with thrombotic microangiopathic hemolytic anemia associated with antiphospholipid antibodies.

Patient No. (reference)	Therapy	Outcome
1 (37)	Steroids	Stabilized but died later of infection
2 (37)	-	Bilateral nephrectomy for hypertension control. Dialysis, transplantation
3 (37)	NR	Died of massive hemorrhage following abdominal surgery
4 (37)	Steroids	Well, but impaired renal function
5 (37)	Heparin+Steroids+Dypiridamol	Well. Renal function improved
6 (37)	PE	Well. Renal function returned promptly to normal
7 (37)	PE	Well. Renal function returned promptly to normal
8 (37)	NR	Well, but impaired renal function
9 (37)	Anticonceptive withdrawal	Spontaneous resolution after ceasing oral contraceptive
10 (37)	NR	Well. Renal function improved
11 (46)	AAS+Infusion of FFP	Well, but impaired renal function
12 (16)	Coumadin+Steroids+PE	Thrombocytopenia unaffected
	Stop PE	Platelet count returned to normal within 5 days after the last plasma exchange.
13 (38)	PE+Dypiridamol+AAS	Well. Renal function improved
14 (29)	Steroids	Platelet count decreased and renal function continued to deteriorate
	PE (FFP)+Cyclo	Her condition dramatically improved
15 (29)	Steroids	Mental status worsened with a right hemiparesis and platelet count decreased
	PE (FFP)+Cyclo	Mental status improved and platelet count began to rise
16 (33)	Steroids	Platelet count decreased
	Heparin	Platelet count increased and creatinine decreased
		Steroids, Azathioprin and warfarin were administered for 2 years without recurrence of thrombotic episodes and normal renal function

Abbreviations: AAS: acetyl-salicylic acid; Cyclo: cyclophosphamide; FFP: fresh-frozen plasma; IVIg: intravenous immunoglobulins; NR: not reported; PS: present series. PE (ALB) or (FFP): plasma exchange using (ALB) albumin or FFP as replacement fluid

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Patient No. (reference)	Therapy	Outcome
17 (33)	Steroids+AAS+PE (FFP and normal saline)	Platelet count increased and creatinine improved and stabilized Discharged on steroids, azathioprin, and AAS. During the next 9 months, hospitalized twice for pulmonary emboli. Over the next 6 months, he had progressive renal insufficiency and finally, he died of <i>Staphylococcus epidermidis</i> endocarditis.
18 (41)	PE (FFP)	Increase in hematocrit and platelet count and improvement of the renal function Discharged on AAS, steroids
19 (72)	AAS+Dipyridamol+PE (FFP)+ Steroids+Cyclo	Hematological improvement was not followed by renal improvement. She died 26 days after admission of catheter-related staphylococcal sepsis
20 (35)	Steroids PE (FFP)	No changes in neurologic status and platelet count decreased Improvement in neurologic status and platelet count began to rise
21 (35)	Steroids+Cyclo+AAS	Patient continued to have thrombocytopenia and episodes of stroke.
22 (35)	Steroids+Cyclo+IVIg PE (FFP)+Vincristine	Platelet count decreased Platelet count increased but hemolytic anemia recurred. Died of acute myocardial infarction (over 1 year)
23 (59)	Steroids PE (FFP and saline)	Clinical status did not improve, platelet count and transaminitis remained abnormal Clinical status improved with normalization of platelet count and liver enzymes
24 (59)	Heparin+Steroids PE (ALB and saline) PE (FFP)	Platelet count decreased with increasing of liver enzymes Marked drop in liver enzymes. Abdominal continued but less severe Improvement with plasma exchange, steroids and anticoagulation. Liver enzymes and platelet count returned to normal. Discharged on warfarin and steroids

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Patient No. (reference)	Therapy	Outcome
25 (40)	PE +Steroids	Clinical status improved markedly. At this point, heparin was initiated. Platelet count increased but creatinine remained elevated. Discharged on steroids and heparin
26 (39)	Infusions of FFP	Thrombocytopenia resolved but renal function remained impaired The patient remained dialysis dependent for 7 months and then there was gradual recovery of renal function
27 (42)	Steroids+Azathioprin	The patient improved progressively over the next 3 weeks
28 (66)	PE (FFP)	Increase in platelet count; upon withdrawal of the PE, thrombocytopenia recurred. When AAS was prescribed (after receiving aCL determination results), the platelet count returned to normal. There was no improvement in renal function, and the patient was discharged on hemodialysis
29 (32)	Infusion of FFP PGE1+Heparin	No significance response was noted Acute renal failure, hypertension and episodic anemia still persisted. Discharged on peritoneal dialysis
30 (54)	Steroids+Cyclo PE (FFP)	Renal function deteriorated over 2-week period Clinical status worsened and patient died of catastrophic APS
31 (60)	Heparin+Steroids+Cyclo IVIg+PE +Steroids	Thrombocytopenia and renal failure remained unchanged Platelet count increased and serum creatinine decreased. Discharged with maintenance immunosuppression consisted of steroids, azathioprin and IVIg every 4 weeks
32 (11)	PE (ALB and FFP)	Platelet count normalized.

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Table W2: Treatment and outcome of patients with thrombotic microangiopathic hemolytic anemia associated with antiphospholipid antibodies.

Patient No. (reference)	Therapy	Outcome
33 (9)		
1 st episode	Steroids+Warfarin	Renal function and mental status returned to baseline. Discharged on warfarin
2 nd episode	Steroids+Warfarin (INR 3.0-3.5)	Transaminitis and microangiopathic hemolytic anemia resolved gradually.
34 (26)	Steroids+Cyclo+AAS	Died of catastrophic APS
35 (58)	NR	NR
36 (4)	PE	Died (21 d): catastrophic APS
37 (23)	Steroids	Platelet count remained persistently low and creatinine began to rise
	PE	Renal function and thrombocytopenia gradually improved
38 (62)	Steroids	Renal function was deteriorating rapidly and dialysis was begun. Discharge on steroids and dipyridamole
39 (53)	PE +Steroids+ AAS+ Dipyridamol+IVIg+Vincristine	In spite of this continuous escalation of treatment, her condition remained refractory, and she died of massive cerebral hemorrhage
40 (53)	PE +Steroids+AAS	Recovery
41 (6)	NR	Died of catastrophic APS
42 (57)	PE (FFP) +Steroids Heparin+ PE +Steroids	Patient remained critically ill After 40 days clinical status stabilized. Discharged on steroids and warfarin. Three weeks later thrombocytopenia developed and neurologic status worsened. IVIg was added to the treatment and the platelet count rose to normal, but the mental status continued to deteriorate. Monthly intravenous cyclo did not improve the patient, and PE without FFP was restarted. Dramatic improvement was noted within 48 hours. Patient responded to repeated PE over 3 years.

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Patient No. (reference)	Therapy	Outcome
43 (3)	Cyclo+Steroids+IVIg+Heparin	Blood pressure remained elevated with worsening of neurologic status. Finally, the patient underwent left nephrectomy because of hematomas that led to severe hypertension. Discharged on steroids.
44 (76)	PE +Heparin+ Steroids+Cyclo	After 3 days rapid neurologic and laboratory improvements were noted Discharged on steroids and coumadin (INR 3-3.5), there was no evidence of recurrent thromboembolism
45 (15)	Infusion of FFP+IVIg PE	Patient developed acute renal failure with severe hypertension Improvement in hematological parameters, but never recovered the renal function and patient received peritoneal dialysis. Seven months later, a relapse was observed with a severe dilated cardiomyopathy and painful acrocyanosis (probable catastrophic APS). PE was restarted with improvement in hematological parameters, but no improvement was noted in cutaneous lesions. Iloprost infusion and low molecular weight heparin were initiated, with no apparent effect, and later on vincristine with a complete remission of the acrocyanosis and an improvement on echocardiographic findings.
PS	Steroids+ PE (ALB) +heparin	Recovery Discharged on tapered steroids and warfarin (INR 2.5-3.5). No further thrombotic events after 12 months of follow-up

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