

Catastrophic antiphospholipid syndrome

Condition for which IVIg use is in exceptional circumstances only

Specific Conditions

- Catastrophic antiphospholipid syndrome

Level of Evidence

Insufficient data (Category 4a)

Intravenous immunoglobulin (IVIg) may be appropriate therapy for catastrophic antiphospholipid syndrome, a term that describes the accelerated form of antiphospholipid syndrome characterised by widespread small vessel thrombosis leading to multiorgan failure. It is not indicated for the treatment of antiphospholipid syndrome in other cases. Please see [Antiphospholipid syndrome \(non-obstetric\) in Chapter 8](#) and Recurrent [foetal loss \(with or without antiphospholipid syndrome\) in Chapter 8](#).

Qualifying criteria for IVIg therapy

A patient will qualify for IVIg when all the following criteria are met:

1. Evidence of rapidly evolving thrombosis involving two or more organs
2. Unequivocal laboratory evidence of antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin antibodies and/or beta 2 glycoprotein I antibodies)
3. Other causes of thrombotic microangiopathy are considered less likely.

Confirmation by histopathology of thrombotic small vessel occlusion in at least one organ or tissue is desirable but should not delay IVIg therapy if indicated.

A single treatment is usually sufficient, based on a dose of 2 g/kg divided over 2–5 days. The potential pro-thrombotic effect of IVIg should be considered in this indication.

Refer to the current product information sheet for further information.

The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.

Bibliography

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