Disseminated Intravascular Coagulation

Synonyms: DIC, consumptive coagulopathy

Description

Usually there is a balance between the clotting and lysis systems; however, in disseminated intravascular coagulation (DIC), the coagulation mechanism (especially thrombin) is activated inappropriately and in a diffuse way. This may lead to thrombosis in the subacute or chronic form but more often haemorrhage occurs as the clotting factors are exhausted. DIC is characterised by evidence of both thrombin and plasmin activation.

The severity of DIC is very variable, ranging from subtle haemostatic dysfunction to severe decompensation and organ failure.[1]

Epidemiology

The condition occurs in response to other pathology rather than as a primary event. There are no predisposing factors in terms of age, sex or race.

Risk factors

Conditions that may be complicated by DIC include:

- Infections, especially septicemia, *Escherichia coli* O157, typhoid fever, Rocky Mountain spotted fever and parasites. The rash of meningococcal septicaemia is classical.
- Malignancy, especially leukaemias.
- Major trauma including crush syndrome and, occasionally, burns.
- Some connective tissue disorders including antiphospholipid syndrome.[2]
- Complications of pregnancy including the placental problem of placental abruption, amniotic fluid embolism, severe hypertension of pregnancy with fulminating pre-eclampsia and HELLP syndrome. A retained dead fetus tends to produce a thrombotic rather than a haemorrhagic state.
- Incompatible blood transfusion.
- Heat stroke.
- Dissecting aortic aneurysm.
- Some snake bites

Presentation

The immediately obvious features are usually those of the underlying condition that has caused DIC, especially if it is acute. In addition there may be large bruises or spontaneous bleeding at venepuncture sites, on the soft palate, legs and the site of trauma.

In subacute or chronic DIC the features may be thrombotic instead with signs of venous thrombosis.

In the acute situation there are many presenting features that may be found:

- Bleeding from at least three unrelated sites is typical and likely sites include:
  - Ears, nose and throat
  - Gastrointestinal tract
  - Respiratory tract
  - Site of venepuncture or IV infusion

- Confusion or disorientation.
- Fever.
- Signs of haemorrhage.
- Signs of adult respiratory distress syndrome (ARDS).
- Skin may show various signs including:
  - Petechiae
  - Purpura
  - Haemorrhagic bullae
  - Acral cyanosis
  - Skin necrosis of lower limbs (purpura fulminans)
  - Signs of thrombosis
  - Localised infarction and gangrene

Investigation
The diagnosis of DIC should include both clinical and laboratory information. If DIC is suspected then clotting screen tests are followed by confirmation:

- Prothrombin time (PT) elevated.
- Activated partial thromboplastin time (aPTT) elevated.
- Platelet counts in DIC are typically low, especially in acute sepsis-associated DIC, but may be increased in malignancy-associated chronic DIC.\[3\]
- Fibrinogen level low.

If two results are positive, diagnosis is possible; if three are positive, it is likely; if all four are positive, it is extremely likely.

Confirmatory tests look for evidence of the simultaneous formation of thrombin and plasmin.

- The D-dimer test gives strong evidence of DIC.
- Fibrin degradation products (FDPs) are helpful but can occur in other conditions such as deep vein thrombosis (DVT) and, in severe disease, they may be negative.
- In acute DIC, PT and aPTT are prolonged, and the platelet count and fibrinogen decrease. D-dimer, FDP, and fibrin monomer levels are elevated.

In subacute or chronic DIC, PT and aPTT may be prolonged or normal. The fibrinogen level is decreased modestly and, in some cases, may be within the reference range or increased. Platelets may be slightly low or normal. D-dimer and FDP are slightly elevated.

Other tests may be useful but do not confirm the diagnosis.

- Thrombin time can be prolonged because of fibrinogen consumption.
- Thrombin-antithrombin complexes in plasma suggest prior thrombin formation.
- Antithrombin levels seem more promising than fibrinogen levels.\[4\]

**Scoring system**\[5\]

The International Society for Thrombosis and Haemostasis (ISTH) DIC scoring system provides objective measurement of DIC. Where DIC is present the scoring system correlates with key clinical observations and outcomes.

**Scoring system for overt DIC** (should only be used if the patient has an underlying disorder known to be associated with overt DIC):

- Order global coagulation tests (prothrombin time, platelet count, fibrinogen, fibrin related marker) and score the test results:
  - Platelet count (>100 x 10^9/L = 0, <100 x 10^9/L = 1, <50 x 10^9/L = 2)
  - Elevated fibrin marker - eg, D-dimer, fibrin degradation products - (no increase = 0, moderate increase = 2, strong increase = 3)
  - Prolonged PT (<3 secs = 0, >3 but <6 secs = 1, >6 secs = 2)
  - Fibrinogen level (>1 g/L = 0, <1 g/L = 1)

- Calculate score:
  - ≥5 - compatible with overt DIC: repeat score daily
  - <5 - suggestive for non-overt DIC: repeat next 1-2 days

**Management**\[6\]

The cornerstone of the management of DIC is treatment of the underlying condition.\[5\] Thus, infection will need antibiotics, and obstetric complications may need intervention.\[6\]

- Transfusion of platelets or plasma (components) in patients with DIC should not primarily be based on laboratory results and should in general be reserved for patients who present with bleeding.
- In patients with DIC and bleeding or at high risk of bleeding (eg, postoperative patients or patients due to undergo an invasive procedure) and a platelet count of <50 x 10^9/L, transfusion of platelets should be considered.
- In bleeding patients with DIC and prolonged PT and aPTT, administration of fresh frozen plasma (FFP) may be useful. It should not be started only on laboratory tests alone but should be considered in those with active bleeding and in those requiring an invasive procedure.
- If transfusion of FFP is not possible in patients with bleeding because of fluid overload, factor concentrates such as prothrombin complex concentrate should be considered (these will only partially correct the defect because they contain only selected factors, whereas DIC involves a global deficiency of coagulation factors).
- Severe hypofibrinogenemia (<1 g/L) that persists despite FFP replacement may be treated with fibrinogen concentrate or cryoprecipitate.
- In cases of DIC where thrombosis predominates, such as arterial or venous thromboembolism, severe purpura fulminans associated with acral ischemia or vascular skin infarction, therapeutic doses of heparin should be considered. If there is a co-existing high risk of bleeding there may be benefits in using continuous infusion of unfractionated heparin (UFH) due to its short half-life and reversibility.
- In critically ill, non-bleeding patients with DIC, prophylaxis for venous thromboembolism with prophylactic doses of heparin or low molecular weight heparin is recommended.
Patients with severe sepsis and DIC may be treated with recombinant human activated protein C by continuous infusion. Patients at high risk of bleeding should not be given recombinant human activated protein C.

In general, patients with DIC should not be treated with antifibrinolytic agents. Patients with DIC that is characterised by a primary hyperfibrinolytic state and who present with severe bleeding could be treated with lysine analogues, such as tranexamic acid.

Complications and prognosis

- DIC is associated with organ failure and it is often fatal condition.[7]
- Organs can be destroyed by infarction, and limb ischaemia can lead to loss of digits or more.
- However, with appropriate care, patients with DIC can have a favourable outcome.[8]
- The prognosis depends mostly upon the underlying condition, but also on the severity of DIC and comorbidity.

Further reading & references

5. Guidelines for the diagnosis and management of disseminated intravascular coagulation, British Committee for Standards in Haematology (January 2009)

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