Disseminated Intravascular Coagulation

Synonyms: consumptive coagulopathy

Description
The International Society of Thrombosis and Haemostasis (ISTH) defines disseminated intravascular coagulation (DIC) as a syndrome characterised by the systemic activation of blood coagulation, which generates intravascular fibrin, leading to thrombosis of small- and medium-sized vessels, and eventually organ dysfunction[1].

Usually there is a balance between the clotting and lysis systems; however, in 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.

Development of acute DIC significantly increases mortality from the underlying condition[2].

Epidemiology[3, 4]
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
Numerous medical conditions may be complicated by DIC including:

- Infections, especially when there is sepsis, which is the most common cause. DIC has been estimated to occur in 30-50% of cases of severe sepsis. Some have reported some degree of coagulopathy, clinical or subclinical, in up to 80%[5]. Gram-negative bacteria are the most common culprits - the rash of meningococcal sepsis is classical. Other infections include Escherichia coli O157, typhoid fever, Rocky Mountain spotted fever and parasites.
- Malignancy, especially leukaemias.
- Major trauma including crush syndrome and, occasionally, burns.
- Complications of pregnancy including the placental problem of placental abruption, amniotic fluid embolism, severe hypertension of pregnancy with fulminating pre-eclampsia and HELLP syndrome.
- Incompatible blood transfusion.
- Transplant rejection
- Severe liver disease
- Pancreatitis
- Heat stroke.
- Dissecting aortic aneurysm.
- Complications after surgery.
- Recreational drugs.
- Some snake bites.
- Some connective tissue disorders including antiphospholipid syndrome[6].

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 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\textsuperscript{[1, 4]}

The diagnosis of DIC should include both clinical and laboratory information. There is no single test which can accurately diagnose DIC alone. Tests have to be repeated to monitor dynamic change.

Findings include the following:

• Platelet counts in DIC are typically low, with a downward trend, especially in acute sepsis-associated DIC. A continuing drop even within normal range may be suspicious.
• Fibrin degradation products including D-dimer are elevated. D-dimer is useful in diagnosis and monitoring of DIC. It is not specific, as a number of conditions cause elevation of D-dimer, but a normal D-dimer excludes DIC as it is highly sensitive\textsuperscript{[7]}.
• Prothrombin time (PT) elevated (prolonged).
• Activated partial thromboplastin time (aPTT) elevated (prolonged).
• Fibrinogen level low, although this may be normal in more than half of cases.

Other findings may include elevated soluble fibrin (SF) and reductions in the levels of natural anticoagulants such as antithrombin (AT) and protein C (PC).

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.

Scoring system\textsuperscript{[1, 4]}

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\textsuperscript{[4]}

The cornerstone of the management of DIC is treatment of the underlying condition. Thus, infection will need antibiotics, and obstetric complications may need intervention. DIC, in many cases, resolves spontaneously once the underlying disorder is treated.

• 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 on laboratory tests alone but should be considered in those with active bleeding and in those requiring an invasive procedure. Initial doses of 15-30 ml/kg are required.
• 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 hypofibrinogenaemia (<1 g/L) that persists despite FFP replacement, or specific fibrinogen deficiencies 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 ischaemia 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.

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 can rapidly lead to organ failure and it is often fatal condition, especially when not identified and treated early.
- Overall mortality from DIC is difficult to establish due to the severity and variety of the underlying conditions. However, it has been estimated that mortality rates for sepsis and severe trauma double if DIC develops.
- 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.

Further reading & references


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