Disseminated Intravascular Coagulation (DIC; Consumptive Coagulopathy) in Infants and Children

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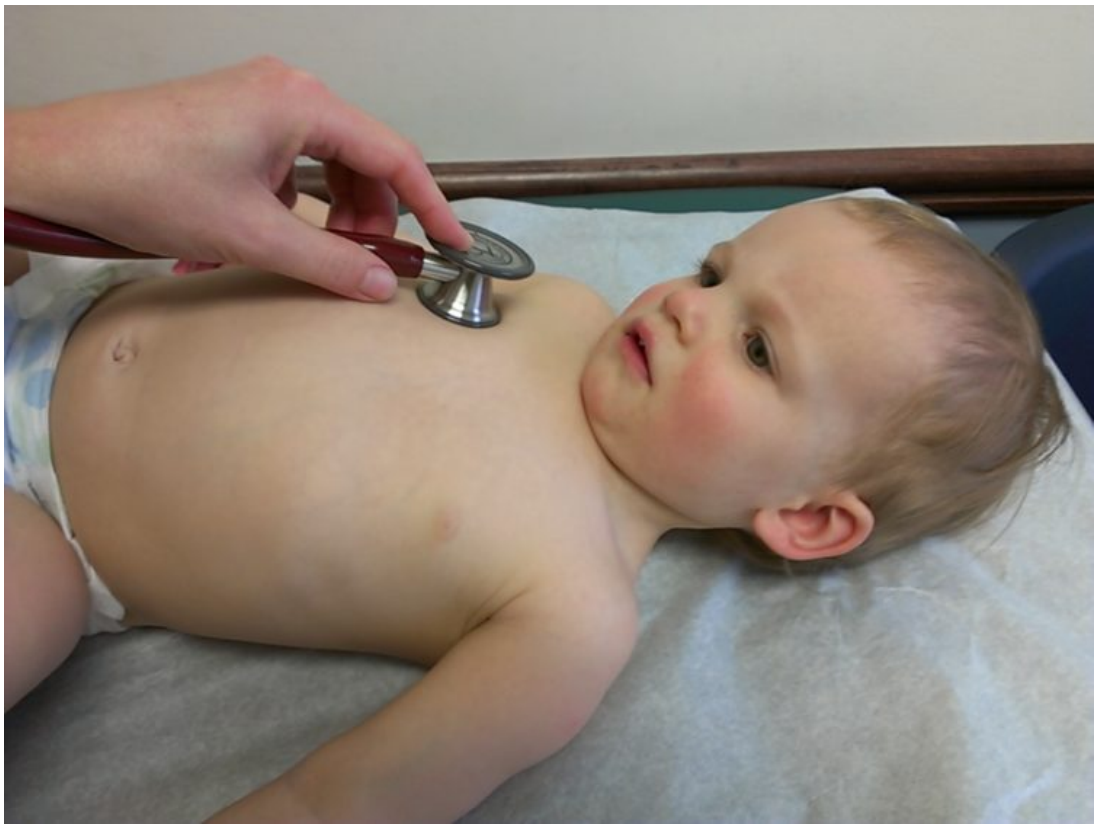
Disseminated Intravascular Coagulation (DIC) is a systemic, life-threatening process associated with both thrombosis and hemorrhage, and it is caused by a wide range of defined disorders. The basic pathology is the activation of coagulation cascade where all the clotting factors get utilized in the formation of an excessive clot, resulting in profuse bleeding. For this reason, it is also known as consumptive coagulopathy. Diagnosis can be made clinically, with the aid of laboratory investigations to confirm the diagnosis.



Definition of Disseminated Intravascular Coagulation

Disseminated Intravascular Coagulation (DIC) is a serious medical disorder in which **multiple clots are formed that can lead to permanent end-organ damage**. Systemic activation of the blood coagulation occurs that results in the generation and the deposition of excess fibrin. This causes the blockage of blood vessels by the thrombi.

It leads towards **multiple organ dysfunctions** as each and every tissue of the body needs a continuous supply of blood to remain alive and functioning. Due to excessive thrombi formation, the platelets and clotting factors become exhausted and depleted. In such a case, any case of minor bleeding will turn into a major bleeding episode.

There are two basic problems in DIC. One is the **occlusion of vessels by the thrombi** and the other is **massive bleeding due to the utilization of all factors in the coagulation cascades**. Both of them are dangerous and can cause very harmful consequences. Once the vicious cycle gets initiated, prompt **therapy is compulsory**; otherwise, the person can die within a few minutes of the onset of DIC.

Types of Disseminated Intravascular Coagulation

It is classified as an acute and chronic form.

The acute form

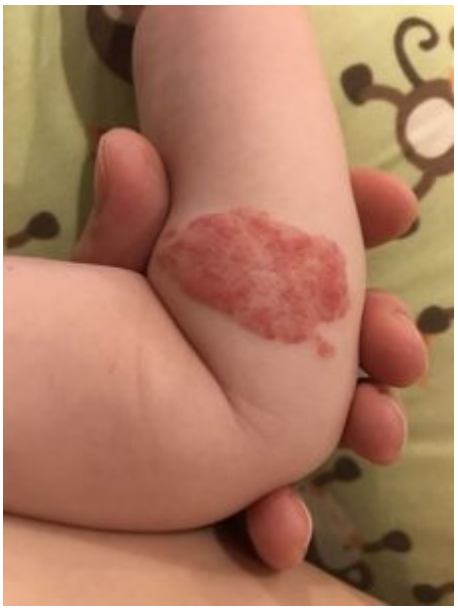
Aggressive intravascular coagulation and consumption of coagulation factors take place. The risk of organ failure is higher.

The chronic form

It is a slow compensated state when the blood is continuously exposed to low levels of stimuli. Permanent organ damage does not result from this. It mostly occurs in the cases of tumors and aortic aneurysms.

Causes of Disseminated Intravascular Coagulation

The most common cause of DIC is **severe sepsis**. It can be due to gram-positive bacteria, gram-negative viruses, parasites, and fungi. Trauma is also a frequent cause of DIC, especially when the SIRS (systemic inflammatory response system) gets initiated. The released inflammatory cytokines activate the coagulation pathways.



- Sepsis
- Severe Infection
- Trauma
- Pancreatitis
- Malignancy
- Transfusion reactions
- Vascular abnormalities
- Hepatic failure
- Toxic reactions
- Surgery
- Burns
- Head injuries
- Rarely: **Antiphospholipid** antibody syndrome
- Large **Hemangioma**

Pathophysiology of Disseminated Intravascular Coagulation

The slow evolving DIC often causes **DVT (Deep Venous Thrombosis), PE (pulmonary embolism), and cardiac vegetations**. The rapid evolving DIC causes **bleeding** as a result of the rapid depletion of **fibrinogen** and clotting factors.

The basic underlying pathology is a disruption of the balance between the Clot formation system and Fibrinolysis in the body tipping in the favor of Clot formation.

The following mechanisms play a role in the causation of disseminated intravascular Coagulation:

When there is endothelial disruption or tissue damage, tissue factor gets exposed. This tissue factor (TF) starts the coagulation cascade and the generation of thrombin. Thrombin converts fibrinogen into active fibrin and clots are formed.

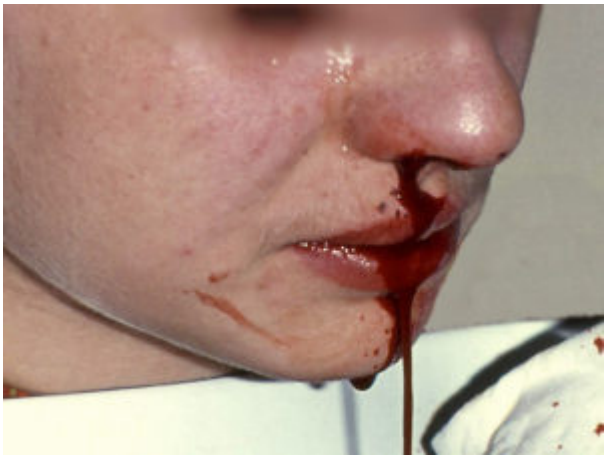
Dysfunctional anticoagulant mechanisms like the depletion of protein C and antithrombin. The fibrinolytic system is depressed as a result of another protein '**plasminogen** activator inhibitor'. The **fibrinolytic** system is important for breaking down the clots, also the release of inflammatory cytokines.

All of the above mechanisms contribute to the following results:

- Increase the platelet formation and aggregation
- Activation of clotting factors
- Formation of excess thrombin
- Thrombin leads to clot formation
- Defective cleavage of clots

Clinical Features of Disseminated Intravascular Coagulation

Following are the **signs and symptoms** of people presenting with Disseminated Intravascular Coagulation:



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- Bleeding from multiple sites in the body
- Bruising
- Toxic looking patient
- Hypotension
- Renal dysfunction
- Respiratory dysfunction
- Hepatic dysfunction
- Central Nervous System Dysfunction
- Deep Venous Thrombosis
- Shock

Diagnosis of Disseminated Intravascular Coagulation

There are three components of the diagnostic workup. The detailed history, physical examination, and laboratory investigations are done.

Physical Examination

Physical Examination is the **most important step**. The treatment is started on the basis of positive physical signs, while the results of the laboratory investigations are awaited. DIC is a life-threatening condition and immediate treatment is started without any delay. The signs to look for during the examination are:

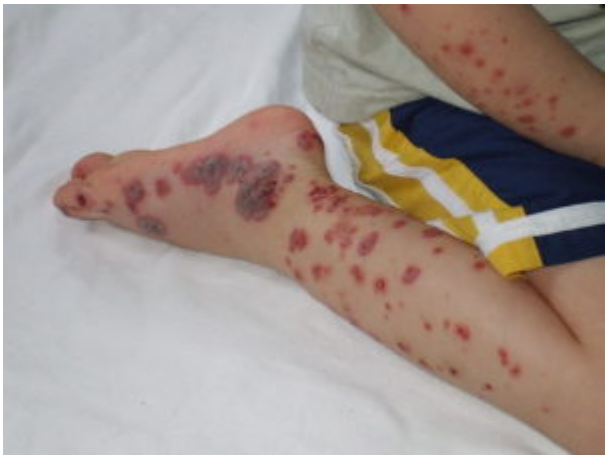

Image: “Purpura2” by Okwikikim. License: CC0 1.0

Signs of hemorrhage

- Signs of localized thrombosis
- Altered consciousness
- Focal neurological deficits
- Hypotension and tachycardia
- Signs of ARDS (Acute Respiratory Distress)
- Hematemesis and haematochezia
- Haematuria
- Oliguria
- Petechiae
- Purpura
- Cyanosis
- Skin necrosis

Laboratory Investigations

Acute DIC

Diagnosis of an acute form of DIC largely depends on the history of the suspected underlying cause (such as sepsis, trauma or malignancy), clinical features, moderate to severe thrombocytopenia (<100,000/microL), specific laboratory findings mentioned below and finding of microangiopathic changes on a blood smear.

- Complete blood count: There is **thrombocytopenia**
- Activated Partial Thromboplastin Time (APTT) and Prothrombin time (PT) are prolonged
- Clotting factors (II, V, and VIII) and Protein C is reduced
- Fibrin Degradation Products (FDP’s) and D-Dimmers are increased

Chronic form of DIC has a lower rate of coagulation factors consumption, but enhancing the synthesis of these factors, thus, there might be the only moderate reduction of platelet count, normal or slightly high plasma fibrinogen, normal PT, and aPTT.

Treatment of Disseminated Intravascular

Coagulation

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Identify and treat the underlying cause.

Administer Fresh Frozen Plasma: The dose is 10–15 ml/kg and it will improve the clotting factor activity by 10–15 %.

Platelet concentrates: 10 ml per kg of platelet concentrates are given to neonates. It will raise the platelet count to 75,000–100,000/ul. The dose is 1 bag/ 5 kg for older children. It is usually indicated when platelet levels reach 10,000-20,000/microL.

Cryoprecipitate: It provides fibrinogen and factor VIII. It is administered when fibrinogen levels drop below 100mg/dL.

Anticoagulant therapy: using Heparin in patients who have or are at risk of venous thrombosis. It is more often used in slow-developing DIC and rarely in rapidly evolving DIC.

The minimum levels required to control the bleeding are:

- Platelets should be > 40,000/ul
- Prothrombin Time < 16 seconds
- Fibrinogen levels > 100 mg/dl

Complications of Disseminated Intravascular Coagulation

- Permanent damage to organs.
- Severe Haemorrhage
- Gangrene
- Cardiac Tamponade
- Stroke
- Shock

- Death

Prognosis of Disseminated Intravascular Coagulation

The prognosis of the disease is **very variable depending on the cause and extent of thrombosis**. An **overall 'poor prognosis'** is associated with DIC. 10–50 % of people die as a result of DIC. Sepsis-related deaths are more common than trauma-related DIC.

References

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Notes