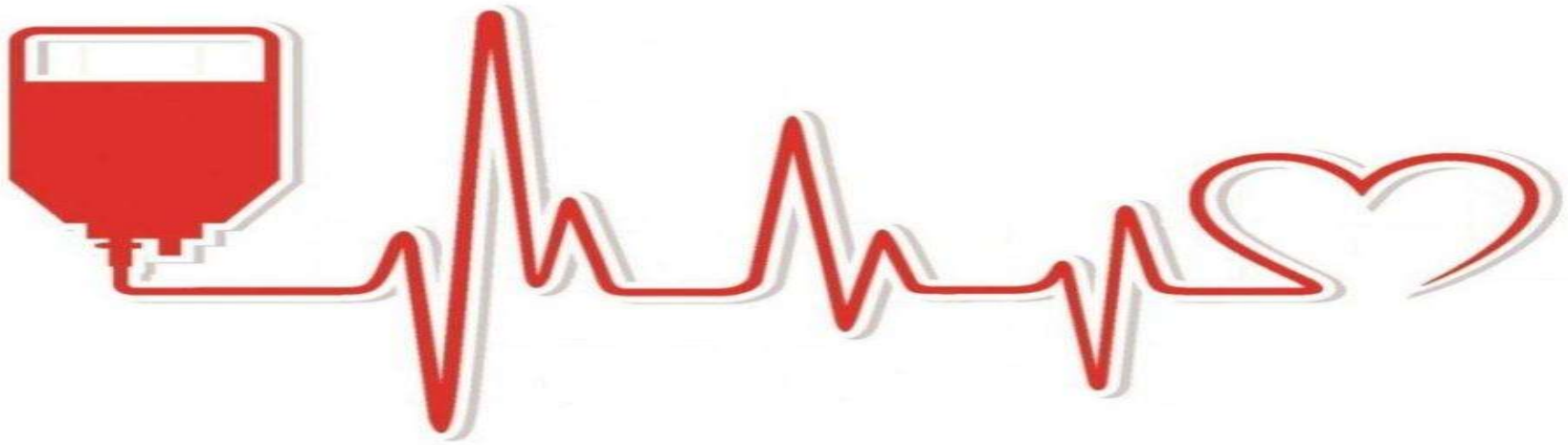
HAEMATOLOGICAL EMERGENCIES



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INTRODUCTION:

A medical emergency is an acute injury or illness

Poses an immediate risk to a person's life or long-term health

Requires immediate medical attention

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INTRODUCTION:

Emergencies in haematological diseases are life and limb threatening disorders

Immediate general supportive and specialized care

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INTRODUCTION:

Depending on the severity of the emergency, and the quality of any treatment given,

May require the involvement of multiple levels of care

From first aiders through emergency medical technicians, paramedics, emergency physicians and anesthesiologists.

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COMMON HAEMATOLOGICAL//ONCOLOGICAL EMERGENCIES

Acute transfusion reactions

TTP

DIC

Cytopenia or Hyperleukocytosis

- Febrile neutropenia
- Acute leukemia with Hyperleukocytosis

Bleeding Diathesis

Crises in sickle cell disease

Metabolic

- Hypercalcemia
- Tumor lysis syndrome

Compressive or Obstructive syndromes

- Spinal Cord compression
- SVC syndrome
- Urologic
- Pericardial
- Central airway

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TRANSFUSION REACTIONS

Transfusions of blood products are associated with several complications,

Can be grouped as immunological or infectious

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ACUTE HEMOLYTIC TRANSFUSION REACTION (AHTR)

also called immediate hemolytic transfusion reaction, is a life-threatening reaction to receiving blood transfusion.

within 24 hours of the transfusion and can be triggered by a few milliliters of blood.

Triggered by pre-formed host antibodies destroying donor red blood cells.

In ABO blood group incompatibility, and is most severe when type A donor blood is given to a type O recipient.

Essential haematology / A.V. Hoffbrand, P.A.H. Moss

Hoffbrand, A. V

View the summary of this work

Bookmark https://trove.nla.gov.au/work/5236388

SERIOUS HAZARDS OF TRANSFUSION (SHOT) DEFINATION OF AHTR

fever and other symptoms/signs of haemolysis within 24 hours of transfusion;

confirmed by one or more of the following:

- **fall of Hb,**
- **rise in lactate dehydrogenase (LDH),**
- **positive direct antiglobulin test (DAT),**
- **positive crossmatch**

SHOT Publications – Articles

2016

Foley K, Poles D, Mistry H, Gray A, Bolton-Maggs PHB. Are the 'rules' for times in set up and duration of red cell transfusion too strict? Transfus Med 2016; 26(3):166-169

EPIDEMIOLOGY

Acute hemolytic transfusion reaction is estimated to occur in 1 in 38,000 to 1 in 70,000 transfusions.

An estimated 41% of ABO-incompatible transfusions result in AHTR.

- *Caligiuri, Michael; Levi, Marcel M.; Kaushansky, Kenneth; Lichtman, Marshall A.; Prchal, Josef; Burns, Linda J.; Press, Oliver W. (2015-12-23). Williams Hematology, 9E. McGraw-Hill Education. ISBN 9780071833004.*

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SIGNS AND SYMPTOMS

fever, rigors (chills).

Mild cases: abdominal, back, flank, or chest pain.

More severe cases: shortness of breath, low blood pressure, haemoglobinuria and may progress to shock and DIC.

In anesthetized or unconscious patients, haematuria may be the first sign of AHTR.

Other: nausea, vomiting, and wheezing

"NHSH Biovigilance Component: Hemovigilance Module Surveiilance Protocol v2.5.2". Retrieved May 1, 2018.

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CAUSES AND PATHOPHYSIOLOGY

Transfusion. 2017 Jun; 57(Suppl 2): 1599–1624.
doi: 10.1111/trf.14168
PMID: 28591471

Supplemental Findings from the National Blood Collection and Utilization Surveys, 2013 and 2015

Mathew R. P. Sapiano,[1,2] Alexandra A. Savinkina,[1,3] Katherine D. Ellingson,[1,4] Kathryn A. Haass,[1] Misha L. Baker,[1,5] Richard A. Henry,[6] James J. Berger,[6] Matthew J. Kuehnert,[1] and Sridhar V. Basavaraju[1]

The most common cause acute hemolytic transfusion reaction is ABO incompability, which is typically due to human error that results in a recipient receiving the incorrect blood product.

Rarely, other blood type incompatibilities can cause AHTR, the most common of which is Kidd antigen incompatibility.

Rh, Kell and Duffy antigen incompatibility have also been implicated in AHTR.

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CAUSES AND PATHOPHYSIOLOGY

Antibodies against A and B blood groups **(isohemagglutinins)** present in the recipient's blood destroy the donor red blood cells

activate the **coagulation cascade (blood clotting system)** via factor XII, which can lead to disseminated intravascular coagulation and kidney damage.

Isohemagglutinins also activate the **complement cascade via C3a and C5a**, which then promote inflammatory cytokine release from WBC.

Inflammatory cytokines include IL-I,IL-6, IL-8, and TNF-alpha, which cause symptoms of low blood pressure, fever, chest pain, nausea, vomiting, and wheezing.

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DIAGNOSIS

Microscopic examination of the recipient's blood and a Direct Antiglobulin Test.

The donor and recipient blood can be re-tested with a type, crossmatch, and antibody screen to determine the cause of the reaction.

"NHSH Biovigilance Component: Hemovigilance Module Surveiilance Protocol v2.5.2". Retrieved May 1, 2018.

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TREATMENT

Initial treatment: discontinuation of the transfusion.

Fluid replacement and close monitoring of vital signs

supportive care,

- include diuretics,
- blood pressure support,
- treatment of disseminated intravascular coagulation (with fresh frozen plasma, cryoprecipitate, and platelet transfusion).

Furosemide is the diuretic of choice

Mannitol.

Dopamine for blood pressure support

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PROGNOSIS

The severity and prognosis depends on the rate of blood administration and the total volume of the transfusion.

Approximately 2% of cases are fatal.

Reactions that begin sooner are typically more severe.

"The 2011 National Blood Collection and Utilization Survey Report"(PDF). Department of Health and Human Services. Retrieved 21 January 2016.

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FEBRILE NONHEMOLYTIC REACTIONS

Why implement universal leukoreduction?

Wafaa Y. Bassuni [a], Morris A. Blajchman [b], May A. Al-Moshary [a]

Due to release of inflammatory chemical signals released by white blood cells in stored donor blood or attack on donor's white blood cells by recipient's antibodies.

This type of reaction occurs in about 7% of transfusions.

Fever is generally short lived and is treated with antipyretics, and transfusions may be finished as long as an acute hemolytic reaction is excluded.

Bassuni, Wafaa Y.; Blajchman, Morris A.; Al-Moshary, May A. (2008). "Why implement universal leukoreduction?". Hematology/Oncology and Stem Cell Therapy. 1 (2): 106–123. doi:10.1016/s1658-3876(08)50042-2.

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FEBRILE NONHEMOLYTIC REACTIONS

Treatment :

Paracetamol

leucoreduction of future transfusions – the filtration of donor white cells from red cell product units

- *"Complications of Transfusion: Transfusion Medicine: Merck Manual Professional". Retrieved 2009-02-09.*

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ALLERGIC TRANSFUSION REACTIONS

caused by IgE anti-allergen antibodies.

histamine is released from mast cells and basophils.

Either IgE antibodies from the donor's or recipient's side can cause the allergic reaction.

common in patients who have allergic conditions such as hay fever.

Patient may feel itchy or having hives but the symptoms are usually mild and can be controlled by stopping the transfusion and giving antihistamines

Laura, Dean (2005). Blood Groups and Red Cell Antigens. Bethesda, United States: National Center for Biotechnology Information. Retrieved 4 October 2017.

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ANAPHYLACTIC REACTIONS

Rare life-threatening allergic conditions **caused by IgA anti-plasma protein antibodies.**

For patients who have selective immunoglobulin A deficiency, the reaction is presumed to be caused by IgA antibodies in the donor's plasma.

Symptoms of fever, wheezing, coughing, shortness of breath, and circulatory shock

Urgent treatment with epinephrine is needed.

Laura, Dean (2005). Blood Groups and Red Cell Antigens. Bethesda, United States: National Center for Biotechnology Information. Retrieved 4 October 2017.

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TRANSFUSION RELATED ACUTE LUNG INJURY (TRALI)

Syndrome that is similar to ARDS,

develops during or within 6 hours of transfusion of a plasma-containing blood product.

Fever, hypotension, shortness of breath, and tachycardia

Diagnosis:

- hypoxemia,
- radiographic evidence of bilateral infiltrates and no evidence of left atrial hypertension (fluid overload).

"NHSN | CDC". www.cdc.gov. 2017-12-29. Retrieved 2018-09-18.

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Korean J Anesthesiol. 2015 Apr; 68(2): 101–105.
Published online 2015 Mar 30. doi: 10.4097/kjae.2015.68.2.101
PMCID: PMC4384395
PMID: 25844126

Transfusion-related acute lung injury; clinical perspectives

Jeongmin Kim[1,2] and Sungwon Na[✉1,2]

Occurs in 15% of the transfused patient with mortality rate of 5 to 10%.

Recipient risk factors includes:

- end-stage liver disease,
- sepsis,
- haematological malignancies,
- ventilated patients.

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TRALI

Antibodies to **human Neutrophil Antigen's (HNA) and Human Leukocyte Antigens (HLA)** have been associated with this type of transfusion reaction.

Donor's antibodies interacting with antigen positive recipient tissue result in release of inflammatory cytokines, resulting in pulmonary capillary leakage.

The treatment is supportive.

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TRANSFUSION ASSOCIATED CIRCULATORY OVERLOAD (TACO)

Common, yet underdiagnosed,

reaction to blood product transfusion

consisting of the new onset or exacerbation of three of the following within 6 hours of cessation of transfusion:

- acute respiratory distress,
- elevated brain natriuretic peptide (BNP),
- elevated central venous pressure (CVP),
- evidence of left heart failure,
- evidence of positive fluid balance, and/or radiographic evidence of pulmonary edema.

Kim, Jeongmin; Na, Sungwon (30 March 2015). "Transfusion-related acute lung injury: clinical perspectives". Korean Journal of Anaesthesiology. 68 (2): 101–105. doi:10.4097/kjae.2015.68.2.101. PMC4384395 PMID25844126

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TRANSFUSION-ASSOCIATED GRAFT VERSUS HOST DISEASE

Frequently occurs in immunodeficient patients where recipient's body failed to eliminate donor's T cells.

Occurs one week after transfusion.

Fever, rash, diarrhoea

Mortality rate is high, with 89.7% of the patients died after 24 days.
Immunosuppressive treatment is the most common way of treatment.

Irradiation and leukoreduction of blood products is necessary for high risk patients for prevent T cells from attacking recipient cells.

Kopolovic, Ilana; Tsubota, Hideki (2015). "A systematic review of transfusion-associated graft-versus-host disease". Blood. 126 (3): 406–414. doi:10.1182/blood-2015-01-620872. PMID 25931584.

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SIGNS AND SYMPTOMS

The clinical presentation is the same as GvHD occurring in other settings, such as bone marrow transplantation. TA-GvHD can develop 2 days to 6 weeks after the transfusion. Typical symptoms include:

fever

erythematous maculopapular rash, which can progress to generalised erythroderma

toxic epidermal necrolysis in extreme cases

hepatomegaly

diarrhea

Other symptoms can include cough, abdominal pain, vomiting, and profuse diarrhea (up to 8 liters/day).

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DIAGNOSIS

A systematic review of transfusion-associated graft-versus-host disease.
(PMID:25931584)

Abstract | Citations | Related Articles | Data | BioEntities | External Links

Kopolovic I[1], Ostro J[1], Tsubota H[2], Lin Y[3], Cserti-Gazdewich CM[4], Messner HA[5], Keir AK[6], DenHollander N[7], Dzik WS, Callum J[3]

Affiliations

Blood [30 Apr 2015, 126(3):406-414]

Type: Review, Journal Article
DOI: 10.1182/blood-2015-01-620872

Laboratory findings include pancytopenia, marrow aplasia, abnormal liver enzymes, and electrolyte imbalance

TA-GvHD can be suspected from a biopsy of the affected skin or liver, and established by HLA analysis of the circulating lymphocytes.

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PREVENTION

Prevention includes gamma irradiation of the lymphocyte-containing blood components such as red blood cells, platelets and granulocytes.

Irradiated blood components should be issued in the following situations:

- **Intrauterine transfusions**
- **Prematurity, low birthweight, or erythroblastosis fetalis in newborns**
- **Congenital immunodeficiencies**
- **Certain hematologic malignancies (e.g. Hodgkin lymphoma)**
- **Patients undergoing hematopoietic stem cell transplantation**
- **Components that are HLA matched, or directed donations from a family member**

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TREATMENT

Treatment is supportive.

No available form of therapy has proven effective in treating TA-GvHD and it is fatal in more than 90% of cases.

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HAEM/ONC EMERGENCIES

Acute transfusion reactions

Thrombotic Thrombocytopenic Purpura

DIC

Cytopenia or Hyperleukocytosis

- Febrile neutropenia
- Acute leukemia with Hyperleukocytosis

Bleeding Diathesis

Crises in sickle cell disease

Metabolic

- Hypercalcemia
- Tumor lysis syndrome

Compressive or Obstructive syndromes

- Spinal Cord compression
- SVC syndrome
- Urologic
- Pericardial
- Central airway

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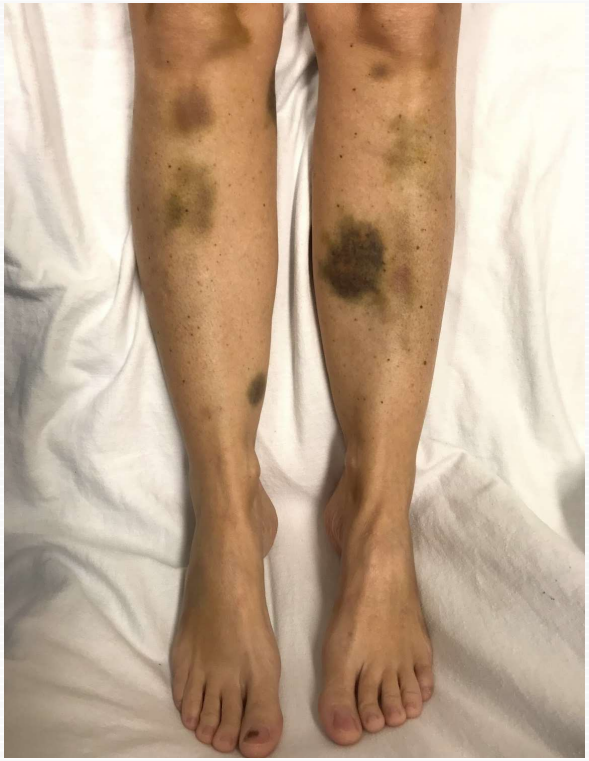
THROMBOTIC THROMBOCYTOPENIC PURPURA (TTP)



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THROMBOTIC THROMBOCYTOPENIC PURPURA (TTP)

Blood disorder that results in blood clots forming in small blood vessels throughout the body. Repeated episodes may occur.

Blood. 2017 May 25;129(21):2836-2846. doi: 10.1182/blood-2016-10-709857. Epub 2017 Apr 17.

Thrombotic thrombocytopenic purpura.

Joly BS[1,2,3], Coppo P[3,4,5], Veyradier A[1,2,3].

Author information

1 Service d'Hématologie Biologique, Hôpital Lariboisière, Assistance Publique-Hôpitaux de Paris, Paris, France.
2 EA3518, Institut Universitaire d'Hématologie Saint Louis, Université Paris Diderot, Paris, France.
3 Centre National de Référence des MicroAngiopathies Thrombotiques and.
4 Service d'Hématologie, Hôpital Saint Antoine, Assistance Publique-Hôpitaux de Paris, Paris, France; and.
5 Université Pierre et Marie Curie, Université Paris 6, Paris, France.

Abstract

Thrombotic thrombocytopenic purpura (TTP) is a rare and life-threatening thrombotic microangiopathy characterized by microangiopathic hemolytic anemia, severe thrombocytopenia, and organ ischemia linked to disseminated microvascular platelet rich-thrombi. TTP is

"Thrombotic thrombocytopenic purpura, acquired". Genetic and Rare Diseases Information Center (GARD) – an NCATS Program. Retrieved 10 October 2018.

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CAUSES

Autoimmune

Genetic

Secondary cause ; Cancer, Bone marrow transplantation, Pregnancy

HIV-1 infection

Medication use:

- Antiviral drugs (acyclovir)
- Certain chemotherapy medications such as gemcitabine and mitomycin C
- Quinine, Oxymorphone
- Quetiapine
- Bevacizumab, Sunitinib
- Platelet aggregation inhibitors (ticlopidine, clopidogrel, and prasugrel)
- Immunosuppressants (ciclosporin, mitomycin, tacrolimus/FK506, interferon-α)
- Hormone altering drugs (estrogens, contraceptives, hormone replacement therapy)

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TTP

Syndrome of target-organ dysfunction caused by marked platelet aggregation in the microcirculation

Explained by an acquired or inherited absence of VWf cleaving protease resulting in ULVWf

- The plasma metalloprotease ADAMTS13 (A Disintegrin And Metalloprotease with ThromboSpondin type 1 motif 13)

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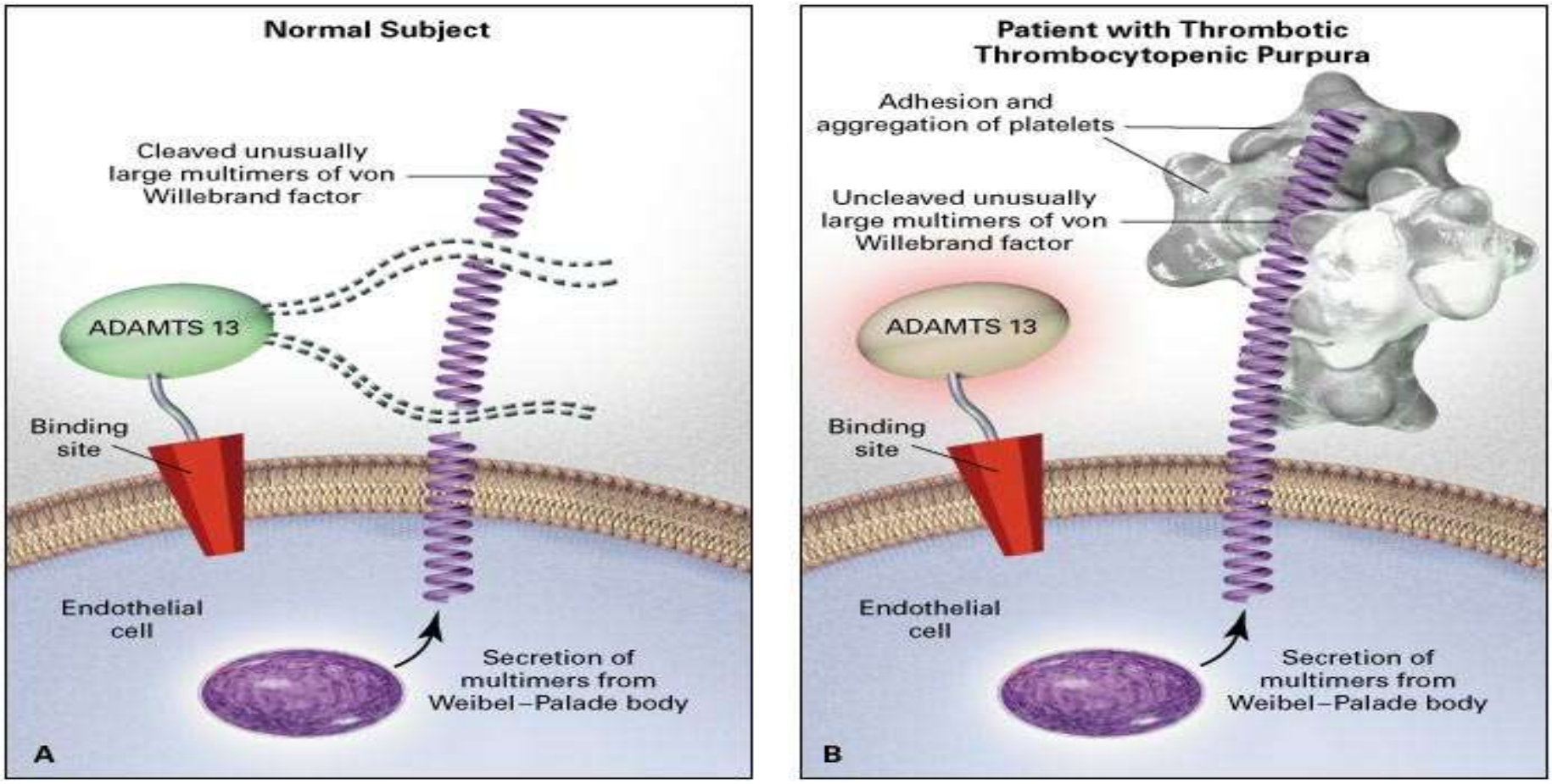
TTP MECHANISM



Known triggers include bacterial infections, certain medications, autoimmune diseases such as lupus, and pregnancy.

The underlying mechanism typically involves antibodies inhibiting the enzyme ADAMTS13.

This results in decreased break down of large multimers of von Willebrand factor (vWF) into smaller units.

Kremer Hovinga, JA; Coppo, P; Lämmle, B; Moake, JL; Miyata, T; Vanhoorelbeke, K (6 April 2017). "Thrombotic thrombocytopenic purpura". Nature Reviews. Disease Primers. 3: 17020. doi:10.1038/nrdp.2017.20. PMID 28382967.

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PRESENTATION

Includes five medical signs. These are:

- Fever
- Changes in mental status
- Thrombocytopenia
- Reduced kidney function
- Haemolytic anaemia (microangiopathic hemolytic anemia)
- High blood pressure (hypertension) may be found on examination.

Allford S, Machin S (2005). "Thrombotic thrombocytopenic purpura". NetDoctor.co.uk.

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DIFFERENTIAL DIAGNOSIS

TTP lead to microangiopathic hemolytic anemia and thrombocytopenia.

This characteristic is shared by two related syndromes, hemolytic-uremic syndrome (HUS) and atypical hemolytic uremic syndrome (aHUS).

George JN (November 2010). "How I treat patients with thrombotic thrombocytopenic purpura: 2010". Blood. 116 (20): 4060–9. doi:10.1182/blood-2010-07-271445. PMID 20686117.

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DIAGNOSIS

low platelet count, low red blood cells due to their breakdown, and often kidneys, heart, and brain dysfunction.

"Thrombotic thrombocytopenic purpura, acquired". Genetic and Rare Diseases Information Center (GARD) – an NCATS Program. Retrieved 10 October 2018.

Joly, BS; Coppo, P; Veyradier, A (25 May 2017). "Thrombotic thrombocytopenic purpura". Blood. 129 (21): 2836–2846. doi:10.1182/blood-2016-10-709857. PMID 28416507.

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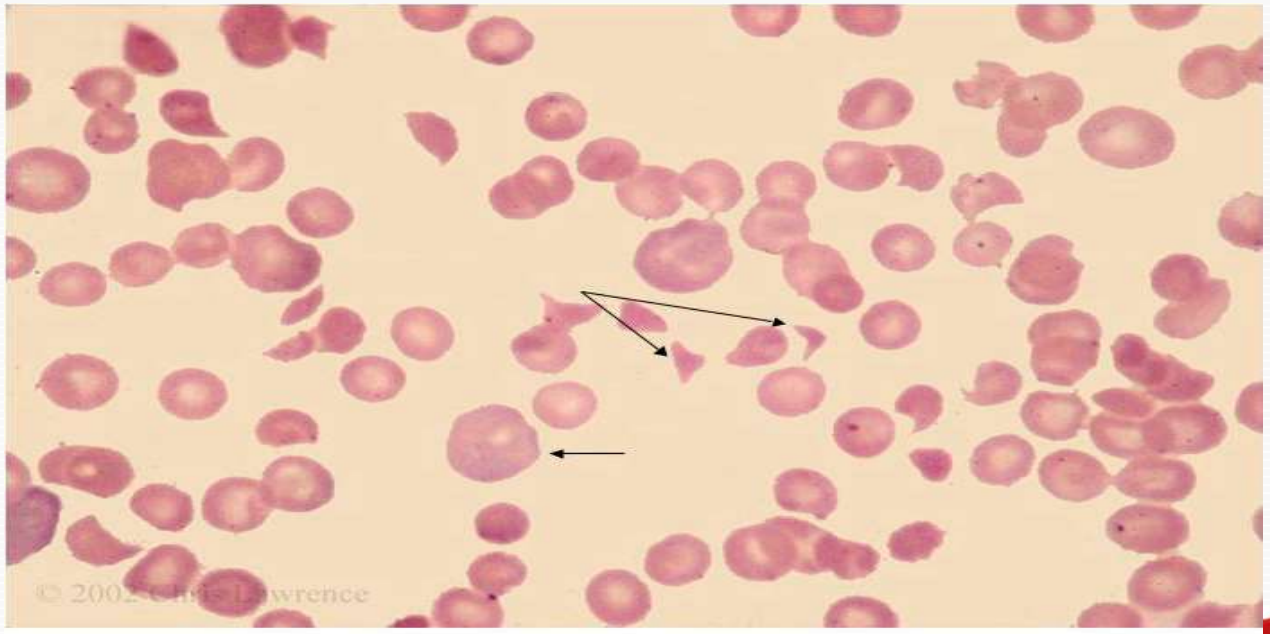
TTP SLIDE



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TREATMENT

high mortality of untreated TTP,

a presumptive diagnosis of TTP is made even when only microangiopathic hemolytic anemia and thrombocytopenia are seen, and therapy is started.

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TREATMENT OPTIONS FOR TTP

Plasma exchange/Apheresis and Plasma infusion

NO platelet transfusion!/Blood transfusion

Immunosuppresion with corticosteroids

Antiplatelet drugs

Splenectomy

Rituximab

N Engl J Med. 1991 Aug 8;325(6):393-7.

Comparison of plasma exchange with plasma infusion in the treatment of thrombotic thrombocytopenic purpura. Canadian Apheresis Study Group.

Rock GA[1], Shumak KH, Buskard NA, Blanchette VS, Kelton JG, Nair RC, Spasoff RA.

Author information

1 Department of Medicine, University of Ottawa, Ont., Canada.

Abstract

BACKGROUND: Thrombotic thrombocytopenic purpura is an uncommon disease with a high mortality rate even with current treatment. The cause of the syndrome and its optimal treatment are unknown. Although both plasma exchange and plasma infusion have been useful treatments, it is not clear which is superior. In this report we describe a prospective randomized trial comparing plasma exchange with plasma infusion for the treatment of thrombotic thrombocytopenic purpura.

Measurements of blood levels of lactate dehydrogenase, platelets, and schistocytes are used to monitor disease progression or remission.

ADAMTS13 activity and inhibitor levels may be measured during follow-up, but in those without symptoms the use of rituximab is not recommended.

Lim W, Vesely SK, George JN (5 March 2015). "The role of rituximab in the management of patients with acquired thrombotic thrombocytopenic purpura". *Blood*. 125 (10): 1526–31. doi:10.1182/blood-2014-10-559211. PMC 4351502. PMID 25573992.

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PROGNOSIS

The mortality rate is around 95% for untreated cases,

Prognosis is reasonably favorable (80–90% survival) for people with idiopathic TTP diagnosed and treated early with plasmapheresis.

Tsai, Han-Mou (February 2006). "Current Concepts in Thrombotic Thrombocytopenic Purpura". Annual Review of Medicine. 57: 419–436. doi:10.1146/annurev.med.57.061804.084505. PMC 2426955. PMID 16409158.

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HAEM/ONC EMERGENCIES

Acute transfusion reactions

TTP

Disseminated Intravascular Coagulation

Cytopenia or Hyperleukocytosis

- Febrile neutropenia
- Acute leukemia with Hyperleukocytosis

Bleeding Diathesis

Crises in sickle cell disease

Metabolic

- Hypercalcemia
- Tumor lysis syndrome

Compressive or Obstructive syndromes

- Spinal Cord compression
- SVC syndrome
- Urologic
- Pericardial
- Central airway

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DISSEMINATED INTRAVASCULAR COAGULATION (DIC)

Condition in which blood clots form throughout the body, blocking small blood vessels.

two main types: acute (rapid onset) and chronic (slow onset).

Loss of hemostasis regulatory mechanisms

Thrombin overproduced ➔ promotes clotting

Fibrin obstructs small vessels

Consumed platelets and factors

"Disseminated Intravascular Coagulation | NHLBI, NIH". www.nhlbi.nih.gov. Retrieved 20 December 2017

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Journal List > Ann Surg > v.229(1); 1999 Jan > PMC1191617

ANNALS OF SURGERY

A Monthly Review of Surgical Science Since 1885

Ann Surg. 1999 Jan; 229(1): 121–127.
doi: 10.1097/00000658-199901000-00016

PMCID: PMC1191617
PMID: 9923809

Disseminated intravascular coagulation and sustained systemic inflammatory response syndrome predict organ dysfunctions after trauma: application of clinical decision analysis.

S Gando, S Nanzaki, and O Kemmotsu

DIC is observed in approximately 1% of academic hospital admissions.

DIC occurs at higher rates in people with bacterial sepsis (83%), severe trauma (31%), and cancer (6.8%).

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CAUSES

Sepsis or severe infection of any kind (infections by nearly all microorganisms can cause DIC, though bacterial infections are the most common): bacterial (Gram-negative and Gram-positive sepsis), viral, fungal, or protozoan infections

Obstetric complications: abruptio placentae, pre-eclampsia or eclampsia, amniotic fluid embolism, retained intrauterine fetal demise, septic abortion, post partum haemorrhage

Massive tissue injury: severe trauma, burns, hyperthermia, rhabdomyolys extensive surgery

Solid tumors and blood cancers (particularly acute promyelocytic leukemia)

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CAUSES

Transfusion reactions (i.e., ABO incompatibility haemolytic reactions)

Severe allergic or toxic reactions (i.e. snake venom)

Giant haemangiomas (Kasabach–Merritt syndrome)

Large aortic aneurysms

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DIC MIMIC

Liver disease, HELLP syndrome, thrombotic thrombocytopenic purpura/Haemolytic uremic syndrome, and malignant hypertension

may mimic DIC but do not occur via the same pathway

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SYMPTOMS

May be lab abnormalities only: no overt clinical signs

Chest pain, shortness of breath, leg pain, problems speaking, or problems moving parts of the body.

As clotting factors and platelets are used up, bleeding.

Blood in the urine, blood in the stool, or bleeding into the skin.

Complications: organ failure.

"Disseminated Intravascular Coagulation | NHLBI, NIH". www.nhlbi.nih.gov. Retrieved 20 December 2017.

^ Jump up to:"Disseminated Intravascular Coagulation (DIC) - Hematology and Oncology". Merck Manuals Professional Edition. September 2016. Retrieved 20 December 2017

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DIAGNOSIS

Typically based on blood tests.

CBC: thrombocytopenia, possible anemia

Blood smear: schistocytes

PT/INR, PTT: prolonged

D-dimer/FDP: elevated

Fibrinogen: diminished

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TREATMENT

Treating the underlying condition.

Transfusions of platelets or fresh frozen plasma can be considered in cases of significant bleeding, or those with a planned invasive procedure.

Cryoprecipitate in those with a low fibrinogen level.

Heparin is rarely used due to the risk of bleeding.

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TREATMENT

Recombinant human activated protein C was previously recommended in those with severe sepsis and DIC, but drotrecogin alfa has been shown to confer no benefit and was withdrawn from the market in 2011.

Recombinant factor VII has been proposed as a "last resort" in those with severe hemorrhage due to obstetric or other causes, but conclusions about its use are still insufficient.

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PROGNOSIS

Between 20% and 50% of patients will die.

DIC with sepsis (infection) has a significantly higher rate of death than DIC associated with trauma.

Becker, Joseph U and Charles R Wira. Disseminated intravascular coagulation at eMedicine, 10 September 2009

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HAEM/ONC EMERGENCIES

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Bleeding Diathesis

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VON WILLEBRAND DISEASE

Mucocutaneous bleeding more likely

Hemarthrosis less likely

Humate F®: FVIII-vWF concentrate

DDAVP only effective in Type 1

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IDIOPATHIC (IMMUNE) THROMBOCYTOPENIC PURPURA (ITP)

Acute in children (age 2 – 6)

- Viral prodrome
- Self-limited: remission in 90%
- Treatment: supportive

Chronic form: adult females

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SIGNS AND SYMPTOMS

Easy bruising, prolonged menses, mucosal bleeding

Petechiae, purpura

Diagnosis: thrombocytopenia on CBC

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TREATMENT

Steroids

If refractory: splenectomy or immunosuppressive therapy

Platelet transfusion ONLY if serious bleeding

Intravenous immunoglobulin G (IVIG) in children with intracranial hemorrhage (rare)

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HEMOPHILIAS

Inherited: sex-linked recessive

Factor VIII: hemophilia A (more common)

Factor IX: hemophilia B

Clinically indistinguishable

PTT elevated in both

CLINICAL SEVERITY

Related to activity level of factor

<1% - severe → frequent spontaneous bleeds

1 – 5% - moderate → occasional spontaneous, lots in trauma

>5% - mild → no spontaneous, excessive in trauma, surgery

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PRESENTATION

Vast majority of disease, present with complications

Most common: intramuscular, intra-articular bleeds

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TREATMENT:
BASED ON COMPLAINT

Minor bleeding: replace 25%

Severe bleeding: replace 50%

Life-threatening: replace 100%

FVIII: 1U/kg ↑ plasma level ~2%

FIX: 1U/kg ↑ plasma level ~1%

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REPLACEMENT FACTORS

Can be recombinant or derived from human plasma

Hemophilia A: mild to moderate → start with DDAVP

- Causes release of vWF and FVIII from endothelial cells

Arthrocentesis rarely indicated

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HAEMO-ONCOLOGICAL COMPLICATIONS

- Hyperviscosity syndrome
- Leukostasis (sludging)
- Neutropenic fever/Febrile neutropenia
- Spinal cord compressions
- Superior vena cava syndrome
- Tumor lysis syndrome
- Hypercalcemia
- Pericardial effusion / tamponade

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HYPERVISCOSITY SYNDROME

- ↑abnormal serum proteins
- Waldenstrom macroglobulinemia
- Multiple myeloma (less common)
- Most common symptoms: neurologic, visual
- May see mucosal or GI bleeding
- CHF from ↑ plasma volume
- Plasmapheresis, exchange

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HYPERLEUKOCYTOSIS /LEUKOSTASIS

WBC sludging in microcirculation

Usually acute leukemia

Can be seen with chronic, NHL

Neurologic symptoms

Can see respiratory failure

Treatment: leukapheresis, hydroxyurea, chemotherapy

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Hematol Oncol. 2016 Jun;34(2):69-78. doi: 10.1002/hon.2292. Epub 2016 Mar 27.

Leukostasis in adult acute hyperleukocytic leukemia: a clinician's digest.

Ali AM[1], Mirrakhimov AE[2], Abboud CN[1], Cashen AF[1].

Author information

1 Washington University School of Medicine, Department of Medicine, St. Louis, MO, USA.
2 University of Kentucky College of Medicine, Department of Medicine, Lexington, KY, USA.

Abstract

Leukostasis is a poorly understood and life-threatening complication of acute hyperleukocytic leukemia. The incidence of hyperleukocytosis and leukostasis differs among various subtypes of leukemias. While the pathophysiology of leukostasis is not fully understood, recent research has elucidated many novel pathways that may have therapeutic implications in the future. Respiratory and neurological compromise represents the classical clinical manifestations of leukostasis. If it is not diagnosed and treated rapidly, the one-week mortality rate is approximately 40%. Targeted induction chemotherapy is an important component of the successful treatment of leukostasis, although other modalities of cytoreduction are being used and investigated.

The incidence and prevalence of hyperleukocytosis and leukostasis varies depending on the form of leukemia.

Hyperleukocytosis is common in chronic myelogenous leukemia and chronic lymphocytic leukemia but leukostasis rarely occurs.

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DIAGNOSIS

White blood counts exceeding 100 x 10^9 / L (100,000 / microL) present symptoms of tissue hypoxia and may signal possible neurological and respiratory distress.

Patients with leukemia need regular neurological and respiratory monitoring when leukocyte counts are approaching 100 x 10^9 / L (100,000 / microL) to decrease chances of tissue hypoxia.

"Hyperleukocytosis and leukostasis in hematologic malignancies". www.uptodate.com. Retrieved 2017-12-12.

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PROGNOSIS

Leukostasis is a high-risk condition

lead to significant complications resulting from occlusion of blood vessels

including transient ischemic attacks and strokes.

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NEUTROPENIC FEVER

Absolute neutrophil count (ANC) = neutrophils + bands

ANC <500 cells/ml

Signs and symptoms: fever

Treatment: IV antibiotics

- Ceftazidime or cefepime ± amino-glycoside or –penem
- Add vancomycin if appropriate

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SPINAL CORD COMPRESSION

Lymphoma, metastasis, primary

C/F: back pain, weakness, numbness, bowel / bladder / sexual dysfunction

Diagnosis: MRI

- CT myelogram if MRI not available

Treatment

- Dexamethasone
- Radiation therapy

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SUPERIOR VENA CAVA SYNDROME

Tumor obstructs SVC ➔ venous hypertension, congestion

Early signs: face edema that improves through day

SOB, cough, chest pain

Distension of chest, neck veins

Cyanosis

Diagnosis: chest CT

Treatment: radiation therapy

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ACUTE TUMOR LYSIS SYNDROME

Usually after chemotherapy

Lysed cells ➔ metabolic changes

Hyperuricemia: renal failure due to renal precipitation

Hyperphosphatemia: lethargy, seizures, renal impair

- Also binds with Ca^{+} → ↓calcium

Hypocalcemia: tetany, arrhythmia

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ACUTE TUMOR LYSIS SYNDROME

Hyperkalemia: muscle weakness, cramps, arrhythmias, heart block ➔ asystole

Acute renal failure: possible hemodialysis; do not alkalinize urine, worsens ↑phosphorus and ↓calcium

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DIAGNOSIS

TLS should be suspected in patients

with large tumor burden who develop acute kidney failure along with hyperuricemia (> 15 mg/dL) or hyperphosphatemia (> 8 mg/dL).

Acute uric acid nephropathy

Urinalysis may show uric acid crystals or amorphous urates.

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HYPERCALCEMIA

BMJ. 2015 Jun 2;350:h2723. doi: 10.1136/bmj.h2723.

The diagnosis and management of hypercalcaemia.

Minisola S[1], Pepe J[2], Piemonte S[2], Cipriani C[2].

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Comment in

Authors' reply to Laurent. [BMJ. 2015]
Problems with the diagnostic algorithm for hypercalcaemia. [BMJ. 2015]

PMID: 26037642 DOI: 10.1136/bmj.h2723

The normal range is 2.1–2.6 mmol/L (8.8–10.7 mg/dL, 4.3–5.2 mEq/L),

Levels greater than 2.6 mmol/L defined as hypercalcemia.

SYMPTOMS

mild increase: no symptoms.

abdominal pain, bone pain, confusion, depression, weakness, kidney stones or an abnormal heart rhythm including cardiac arrest.

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DIAGNOSIS

By either a increased corrected calcium or ionized calcium level and be confirmed after a week.

Specific changes, such as a shortened QT interval and prolonged PR interval, may be seen on an electrocardiogram (ECG).

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TREATMENT

Intravenous fluids, furosemide, calcitonin or pamidronate in addition to treating the underlying cause.

In those with very high levels, hospitalization and Haemodialysis

In those with vitamin D toxicity, steroids may be useful.

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TAKE AWAY MESSAGE

Emergencies in haematological diseases are life and limb threatening disorders needs immediate general supportive and specialized care

The first step in managing any transfusion reaction is to stop the transfusion

Hemarthrosis in hemophiliacs: factor replacement, never arthrocentesis

NO PLATELET TRANSFUSION in patients with TTP, HUS

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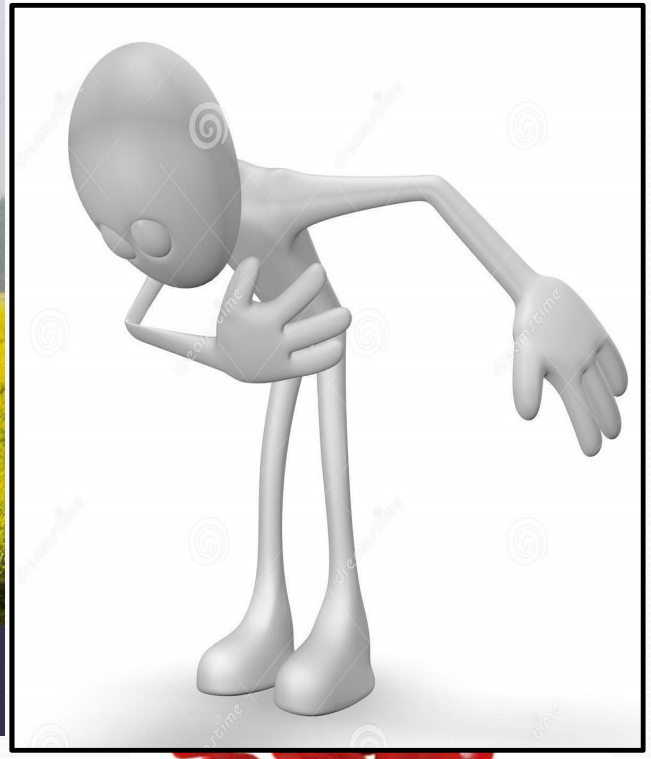
THANKS



Millions of best wishes to you all

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