

Hyperviscosity I.

Common hematological emergency, defined as increase in whole blood viscosity as a result of an increase in:

- Red cells
- White cells
- Plasmatic components, (Ig)

Other additional conditions:

- temperature,
- hydratation status,
- size of cell (CLL vs. AML)

Hyperviscosity II.

Hyperviscosity cause tissue hypoperfusion and consequent complications and clinical features coming from multiple organ hypoxemia:

- Central nervous system disturbances
- Renal impairment
- Respiration insufficiency
- Myocardial infarction

Etc....

Clinicaly relevant hyperviscosity syndrom must be managed inpatient settings and urgent treatment in specialised hematological centre is warranted.

Red cells:

Polycythaemia vera - increased red cell volume,
Hb > 180 gr/l.

Clinical features:

lethargy, headache, hypertension, arterial thromboses
(MI, visual loss – central retinal artery occlusion).

Emergency treatment: = Isovolaemic venesection.

removal of 500mL blood volume from large vein with
simultaneous replacement into another vein of 500mL
0,9% saline.

If present or not emergent – **Erythrocytaferesis**

On cell separator machine until syndrom resolves or until
hematocrit decrease bellow 45%

White cells = Hyperleukocytosis

Acute leukemia – AML, ALL

Chronic leukemia – CML, CLL

Typicaly high blast cell numbers circulating in peripheral blood at presentation/diagnosis.

Leukocytes are sludging in capillaries causing organ damage

More common in AML and blast crises of CML (size of blasts!)

Tumor burden... WBC > 50-100 x10⁹/L

Pulmonary haemorrhage and haemoptysis may occur

White cells = Hyperleukocytosis

Clinical features:

- **pulmonary leucostasis** – cough, dyspnoea, respiratory distress syndrom, tachypnoea, hypoxaemia, diffuse interstitial infiltrate on CRX
Differentiation from bacterial or fungal pneumoia may be difficult...
- **cerebral leucostasis** – encefalopathy, confusion, decreased conscious level, isolated cranial nerve paresis
- visual loss (arteria centralis retinae hemmorrhage/trombosis)
- renal impariment – lab, oliguria...

White cells = Hyperleukocytosis

Emergency treatment:

Anaemia may protect patient from hyperviscosity. Transfusion of RBCs to correct anaemia may initiate leucostasis and worsen the clinical status

Leucapheresis on cell separator machine immediately, usually 2 hour procedure

Continue leucoapheresis daily until leucostasis syndrom is resolved or until $WBC < 50 \times 10^9/L$

Start chemotherapy as soon as criteria allow.

Leucoaferesis is some kind of emergency and bridge to the chemotherapy.

Dubling time of acute leukemia could be faster then leucoapheresis

Plasma components - Immunoglobulins

group of diseases characterized by monoclonal proliferation of the cells of B-lymphoid line, secreting immunoglobulins:

Monoclonal protein in plasma

Multiple myeloma - IgG/IgA paraprotein

Waldenstrom macroglobulinaemia - IgM

Clinical features:

Neurological symptoms: sleepiness, headache, dizziness, coma

Bleeding: interference of Ig with clotting factors

Myelomic nephropathy: Accumulation of Bence-Jones proteinuria in renal tubules, hypercalcaemia.

Plasma components - Immunoglobulins

Hyperviscosity usually develop when total protein in blood is >110 gr/L

Emergency treatment = Plasmapheresis on cell separator machine with the aim of 1,5-2,0 x blood volume exchange.

Repeating daily until symptoms are resolved or total protein < 110 g/L.

After stabilization specific chemotherapy should be initiated

Neutropenic fever

One of the commonest hemato-oncological emergencies.

Definition:

- Presence of symptoms or signs of infection in a patient with absolute neutrophil count $< 0,5 \times 10^9/L$.
- Fever – 1x TT $> 38,3$ C, 2x in 1 hour 38,0 C
- Neutrophils are the natural barrier against bacterial and fungal agents

Neutropenic fever - specialities

Neutropenia=

- Limited ability to produce inflammatory infiltrate
- infections do not behave as usual:
no absces but flegmonous infection
- Pneumonia without pneumonia at CRX
clinical signs (cough, auscultation,CRP) and no finding on
CRX (HRCT)
- Occasionally the fever is the only sign of infekction

Neutropenic sepsis

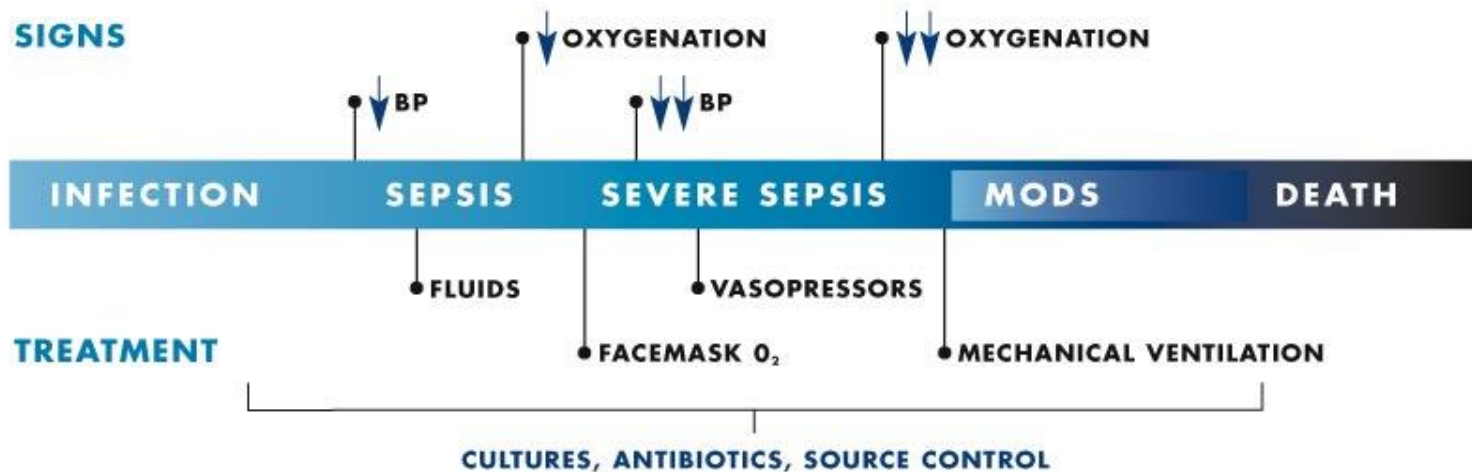
Similarly to polytraumas, MI and stroke, speed and accurate management strongly influence the patient survival

Progression of sepsis to acute organ failure directly affects mortality:

- cca 15% mortality in sepsis **without** organ failure
- cca 70% mortality in **≥3** selhávajících orgánů

Neutropenic sepsis

CONTINUUM OF THE INFECTIOUS PROCESS



1.SIRS = systemic inflammatory response syndrome

Infection/
Trauma

SIRS

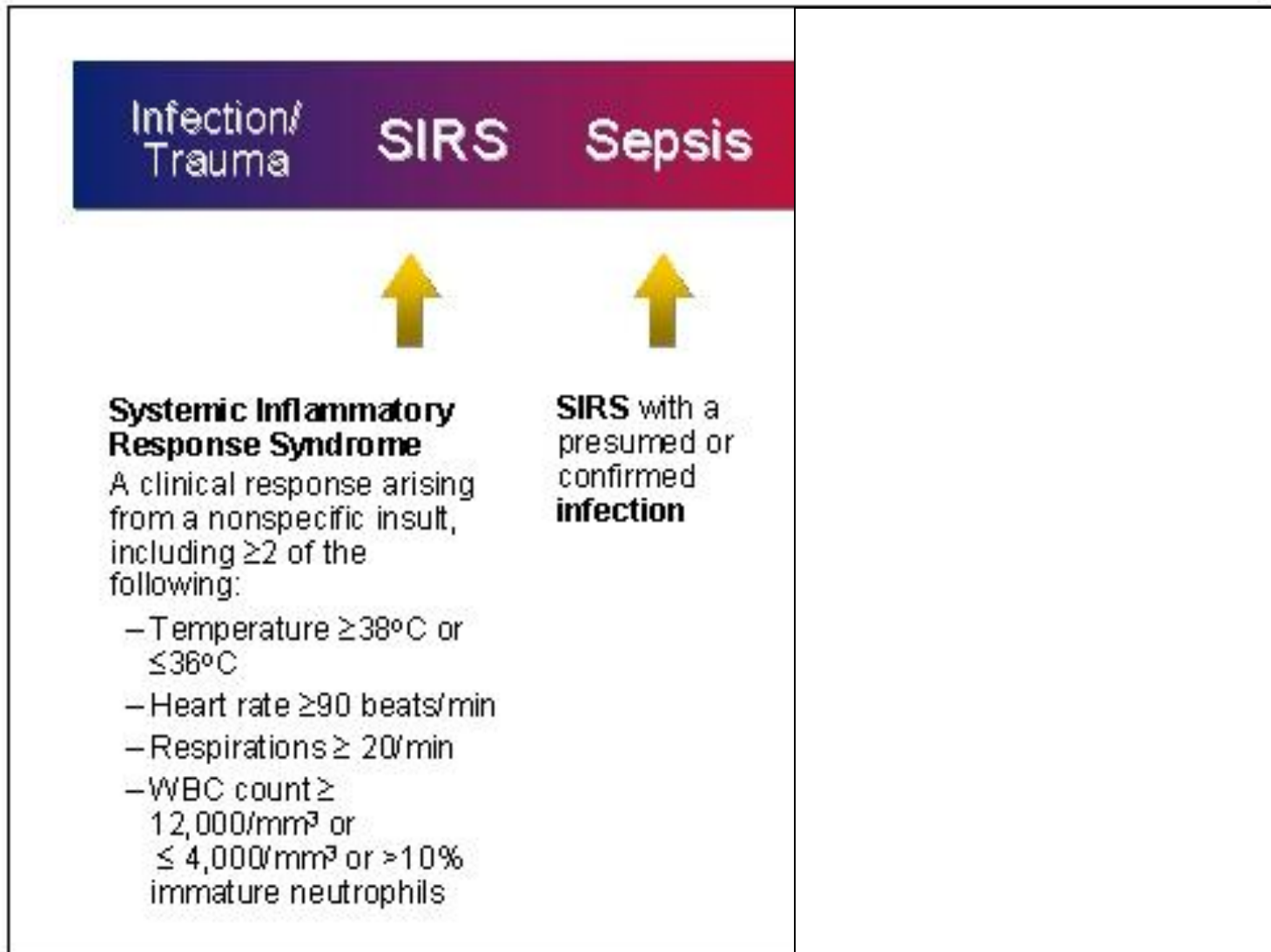


Systemic Inflammatory Response Syndrome

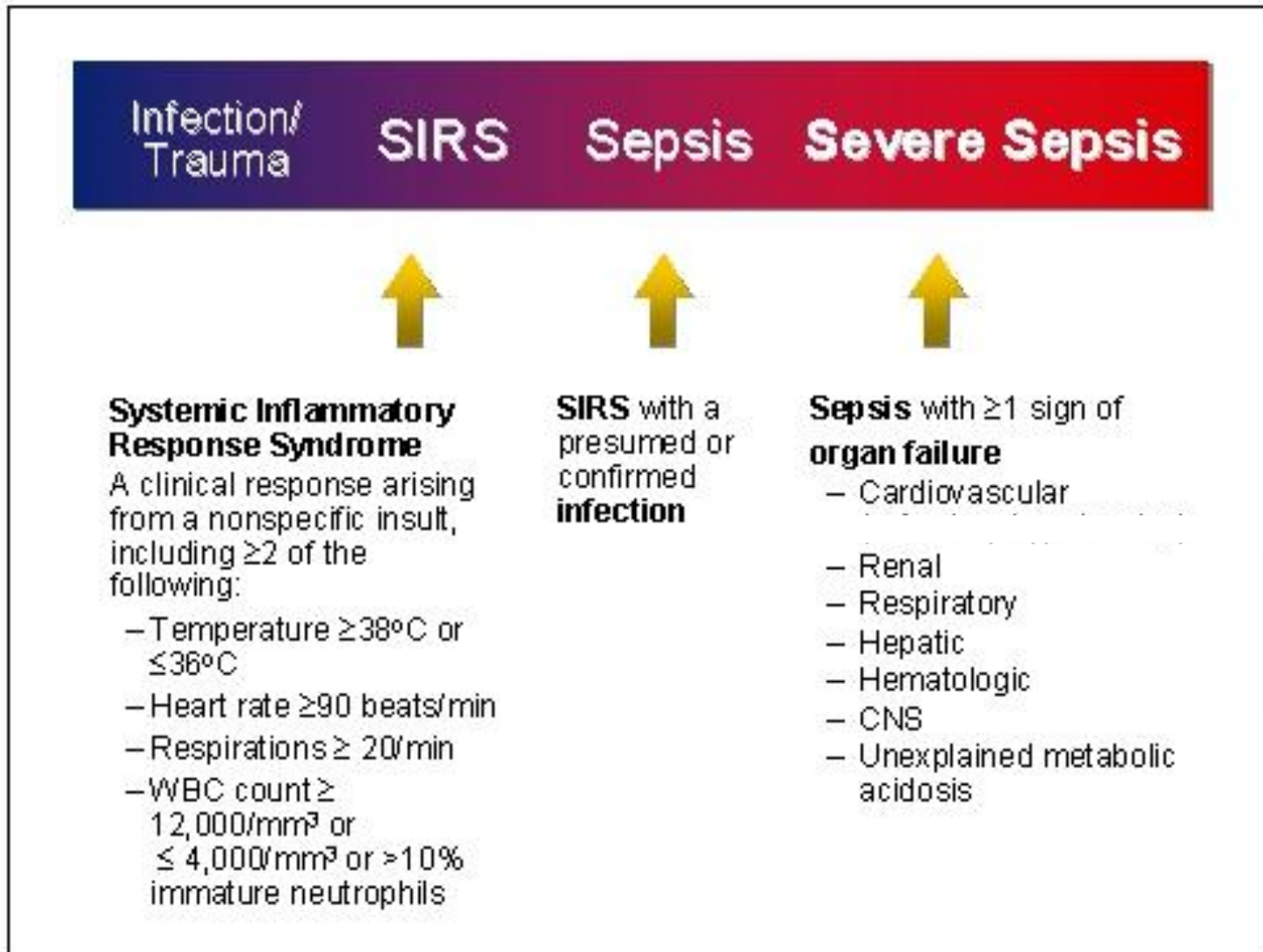
A clinical response arising from a nonspecific insult, including ≥ 2 of the following:

- Temperature $\geq 38^{\circ}\text{C}$ or $\leq 36^{\circ}\text{C}$
- Heart rate ≥ 90 beats/min
- Respirations ≥ 20 /min
- WBC count $\geq 12,000/\text{mm}^3$ or $\leq 4,000/\text{mm}^3$ or $>10\%$ immature neutrophils

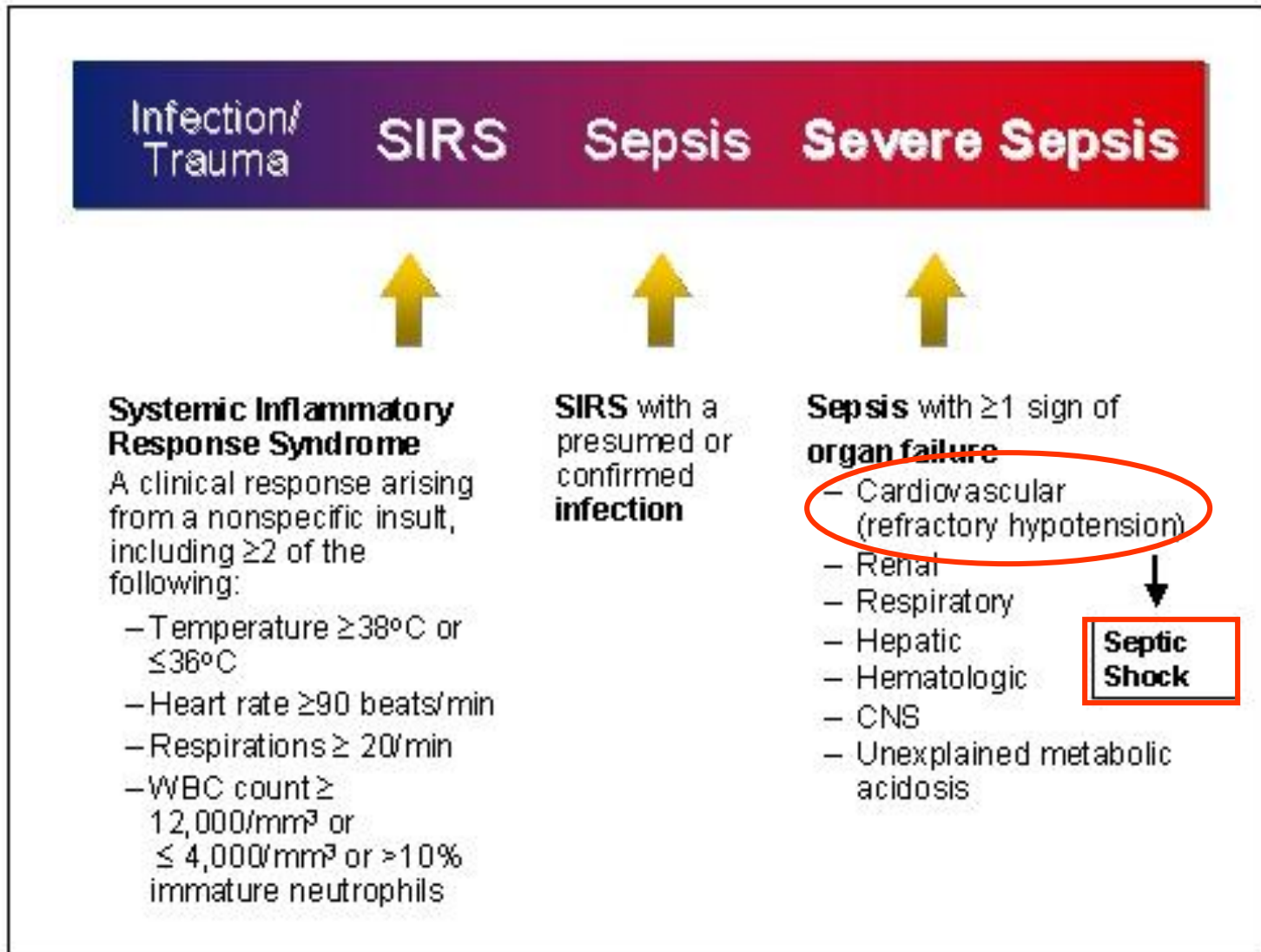
2. SEPSIS = SIRS + presumed / proven infection



3. SEVERE SEPSIS = sepsis + ≥ 1 organ failure



4. SEPTIC SHOCK = severe sepsis + hypotension not responding to parenteral volume resuscitation



Patient risk stratification

- **Neutropenia.**
 - severity of neutropenia:
($<1 \times 10^9$ $<0.5 \times 10^9$ $<0.1 \times 10^9$)
 - length:
(7.... 30 days)
- **Basic diagnose:**
(solid tumor....acute leukemia....HSCT)
- **Status of the disease:**
(remission.....progression)
- ICU patient with multiple infections and organ failure

The Severe Sepsis Resuscitation Bundle

Management of severe sepsis:

6 interventions in, first 6 hours from diagnosis

Decrease in mortality in 35%

The Severe Sepsis Resuscitation Bundle

First of all, patient must be provided by 2 save cannulas

1. Plasma lactate level

Excellent marker of tissue/organ hypoperfussion and hypoxia

Usually increasing earlier then patient becomes hypotensive.

The Severe Sepsis Resuscitation Bundle

2. Blood sampling for hemoculture

before starting any antibiotic therapy

If any catheter is present, hemocultures should be drawn from the device as well to exclude catheter infection

The Severe Sepsis Resuscitation Bundle

3. Start proper parenteral antibiotics

with broad spectrum (antipseudomonal activity!) at the very latest 1 hour from the diagnosis of severe sepsis

Antipseudomonal penicillin with clavulanic acid (Tazocin)
In combination with aminoglycoside (Amikin, Gentamycin)

Alternative – carbapenems

Any restrictive speculation could be fatal – we start with broad spectrum ATB, waiting for cultures and after defining the exact pathogen, we can deescalate ATB.

The Severe Sepsis Resuscitation Bundle

4. Presence of hypotension

(SBP < 90 mmHg nebo MAP < 65 mmHg)

Urgently start **parenteral fluid challenge**

= 1000 ml of crystalloid or 500ml of colloid solution
over 30 minutes

Close monitoring of vital signs

Assess effect: BP, pulse....

The Severe Sepsis Resuscitation Bundle

5. Hypotension persists after fluid challenge

Contact ICU specialist, reserve ICU bed

Central venous catheter, further fluids and CVP (>10 cm H₂O)

Start vasopressors (norepinephrin), invasive blood pressure
goal = MAP > 65 mmHg

Close monitoring of diuresis

The Severe Sepsis Resuscitation Bundle

6. Persisting lactate level despite of fluid and vasopressor optimization

Signaling high mortality...

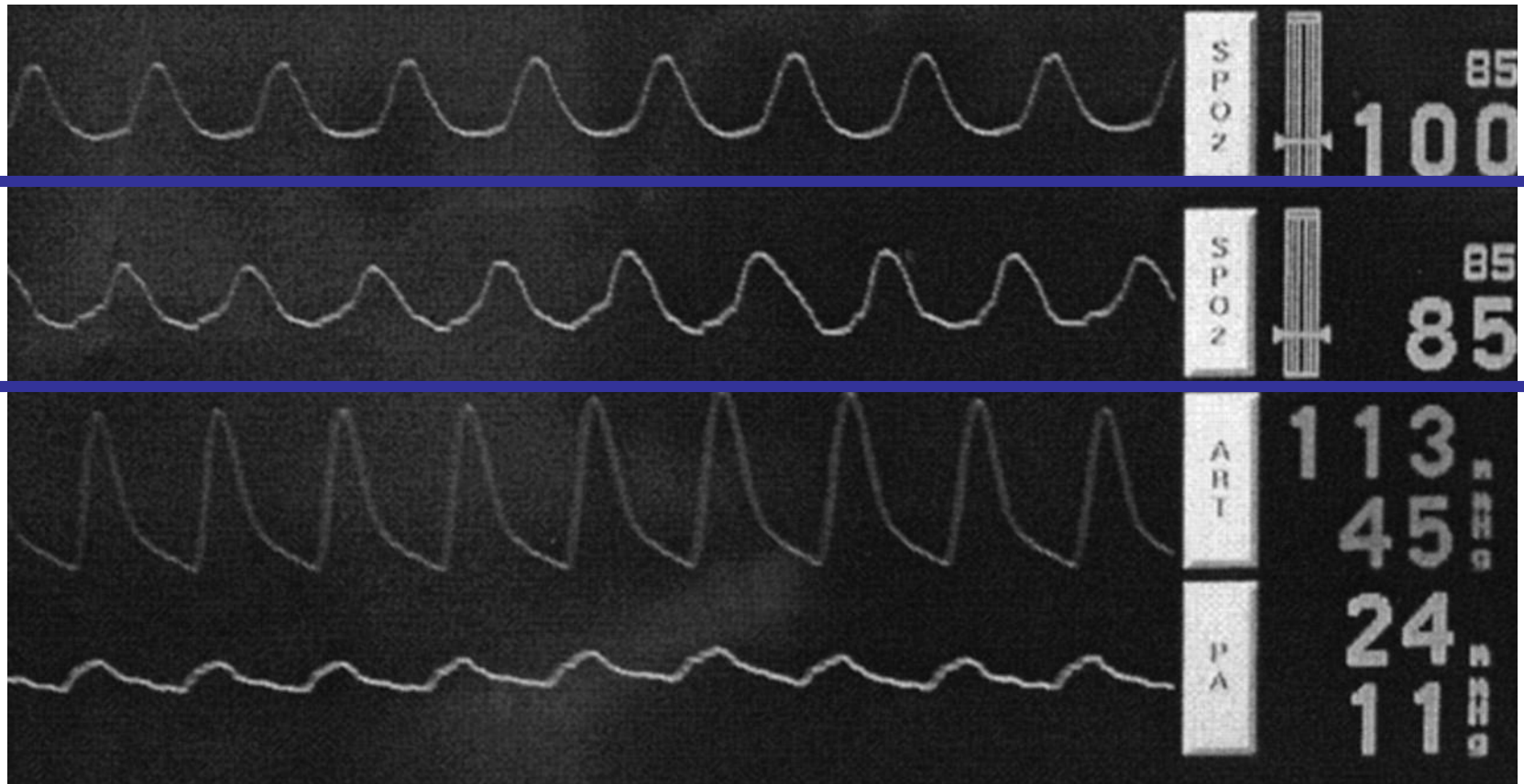
Continue with fluid overload to CVP 15-20 (pulmonary edema)

Correct anaemia, Htc more 30%

Start inotropic agents (dobutamin)

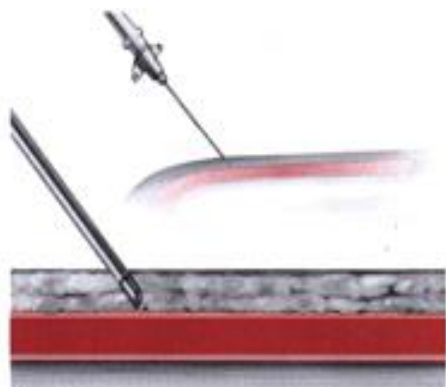
IX. Neinvazivní monitorace - poznámky

Pulzní oxymetrie

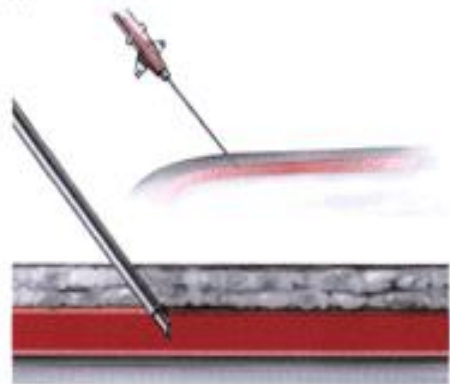




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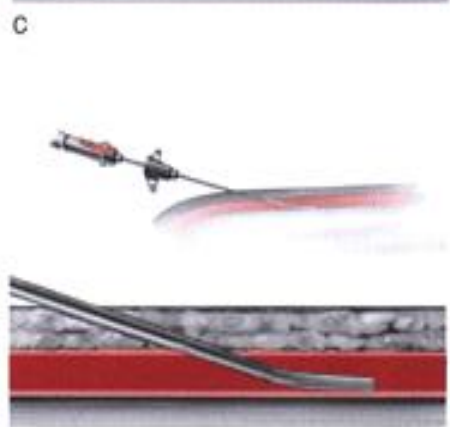
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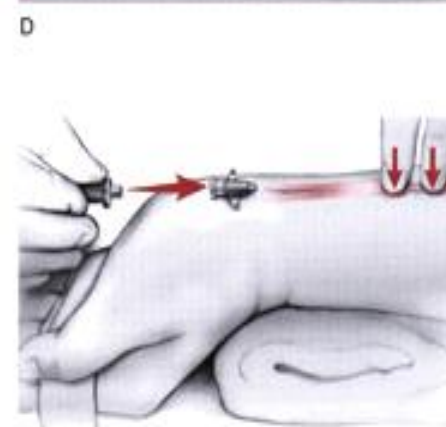
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