



A case of late onset endothelial complication after hemopoietic stem cell transplantation

Spunti di discussione

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Spunti

- Tossicità cumulata
- Esordio tardivo
- Manifestazione clinica atipica e improvvisa
- Diagnostica rapida e invasiva
- Trattamento precoce e tempestivo

Esordio tardivo

Table 1. Clinical criteria for veno-occlusive disease.

Modified seattle criteria (Shulman & Hinterberger, 1992)	Baltimore criteria (Jones <i>et al</i> , 1987)
Two of the following criteria must be present within 20 d of transplant:	Bilirubin must be $>34.2 \mu\text{mol/l}$ (2 mg/dl) within 21 d of transplant and two of the following criteria must be present:
Bilirubin $>34.2 \mu\text{mol/l}$ (2 mg/dl)	Hepatomegaly
Hepatomegaly or right upper quadrant pain	Ascites
Weight gain ($>2\%$ from pre-transplant weight)	Weight gain ($>5\%$ from pre-transplant weight)

Table 2. New EBMT criteria for SOS/VOD diagnosis in adults

<i>Classical SOS/VOD</i> In the first 21 days after HSCT	<i>Late onset SOS/VOD</i> > 21 Days after HSCT
Bilirubin ≥ 2 mg/dL and two of the following criteria must be present:	Classical VOD/SOS beyond day 21
Painful hepatomegaly	OR
Weight gain $> 5\%$	Histologically proven SOS/VOD
Ascites	OR
	Two or more of the following criteria must be present:
	Bilirubin ≥ 2 mg/dL (or $34 \mu\text{mol/L}$)
	Painful hepatomegaly
	Weight gain $> 5\%$
	Ascites
	AND Hemodynamical or/and ultrasound evidence of SOS/VOD

Abbreviations: EBMT = European Society for Blood and Marrow Transplantation; SOS = sinusoidal obstruction syndrome; VOD = veno-occlusive disease. These symptoms/signs should not be attributable to other causes.

Manifestazione clinica atipica e improvvisa

Esordio atipico:
+33

Bilirubina 1,2 mg/dl
Ascite, Creatinina 3
mg /dl
Transaminasi > 5
volte

I nuovi criteri clinici EBMT descrivono la SOS a esordio tardivo

Late onset of VOD:

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Therefore,

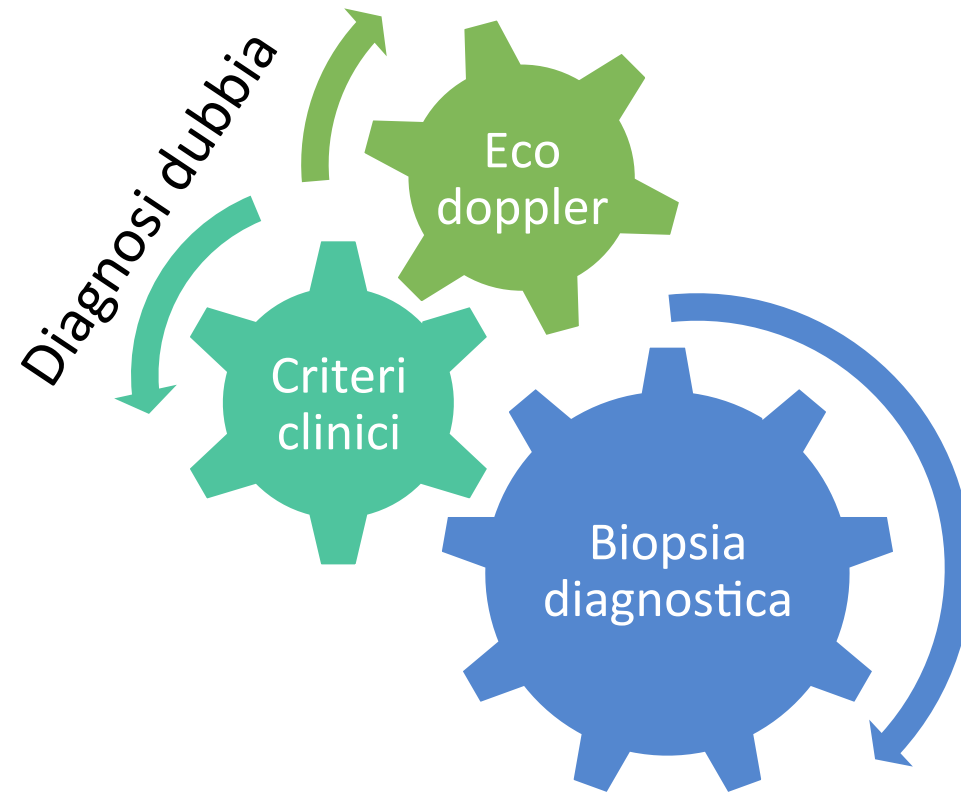
hyperbilirubinemia should no longer be mandatory in late onset SOS/VOD, and the diagnosis of late onset SOS/VOD may be made if patients fulfill a less stringent version of the Baltimore criteria, i.e., at least two of the following:

- bilirubin \geq 2 mg/dL,
- Painful hepatomegaly,
- weight gain > 5% or ascites.

However, hemodynamical and/or ultrasound evidence of SOS/VOD (hepatomegaly, ascites, decrease in velocity or reversal of the portal flow...) is mandatory in addition to these criteria.

Finally, although transjugular liver biopsy is invasive and difficult to perform, histological evidence of SOS/VOD remains the gold standard (but not mandatory) for the diagnosis.

Diagnostica rapida e invasiva



Decisione terapeutica

Table 3. New EBMT criteria for severity grading of a suspected SOS/VOD in adults

	Mild ^a	Moderate ^a	Severe	Very severe - MOD/MOF ^b
Time since first clinical symptoms of SOS/VOD ^c	>7 Days	5-7 Days	≤4 Days	Any time
Bilirubin (mg/dL)	≥ 2 and < 3	≥ 3 and < 5	≥ 5 and < 8	≥ 8
Bilirubin (μmol/L)	≥ 34 and < 51	≥ 51 and < 85	≥ 85 and < 136	≥ 136
Bilirubin kinetics			Doubling within 48 h	
Transaminases	≤ 2 × normal	> 2 and ≤ 5 × normal	> 5 and ≤ 8 × normal	> 8 × Normal
Weight increase	< 5%	≥ 5% and < 10%	≥ 5% and < 10%	≥ 10%
Renal function	< 1.2 × baseline at transplant	≥ 1.2 and < 1.5 × baseline at transplant	≥ 1.5 and < 2 × baseline at transplant	≥ 2 × baseline at transplant or others signs of MOD/MOF

- SOS/VOD severa: mortalità > 80%
- Il trattamento precoce con defibrotide, visto anche il coinvolgimento renale, ha consentito di avere un outcome clinico positivo.

A landscape photograph featuring a vibrant rainbow arching across a blue sky with scattered white clouds. Below the rainbow is a dark green forest line, and in the foreground, a golden-yellow field is illuminated by sunlight, creating long shadows. The overall scene is serene and natural.

TROMBOMODULINA

PAI-1

VCAM1

ICAM 1

BIOMARKERS ?

vWF

L-FICOLINA

ANGIOPOIETINA2

AC. IALURONICO

