State of the Art Safety Standards in RA
THE EUROPEAN SOCIETY OF REGIONAL ANAESTHESIA & PAIN THERAPY

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ESRA ITALIA

www.esraitalia.it

ESRA MEETING
ANNUAL UPDATE

1 day, 1 programme, 3 cities

NAPOLI, 13 APRILE 2024

Presidente:
Giuseppe Servillo

Responsabile scientifico:
Giuseppe Lubrano
Antonio Coviello

Update in anestesia ostetrica

Maria Grazia Frigo
UOSD Anestesia e Rianimazione Ostetrica
Ospedale Isola TiberinaGemelli Isola
Resp SIAARTI Sez Cure Materno Infantili

La frase più pericolosa in assoluto è:

La frase | "Abbiamo sempre fatto così". ıto è:

Grace Murray Hopper tto così".

Grace Murray Hopper

Deficit FVII

↓

ALR vs AG

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Factor VII is a vitamin K dependent plasma protein

DEFICIT F-VII

- Rare bleeding disorder
- Mutation of the gene lacated on chromosome 13
- Autosomal recessive
- 1: 300000-500000
- Heterozygous form 1:350

CLASSIFICATION

- ❑SEVERE: FVII <10% risk of spontaneous major bleeding
- ❑MODERATE: FVII 10-20% risk of mild spontaneous or triggered bleeding
- ❑MILD: FVII 20-50%. Mostly asymptomatic disease

Nevertheless,.....FVII activity level does not always correlate with bleeding severity

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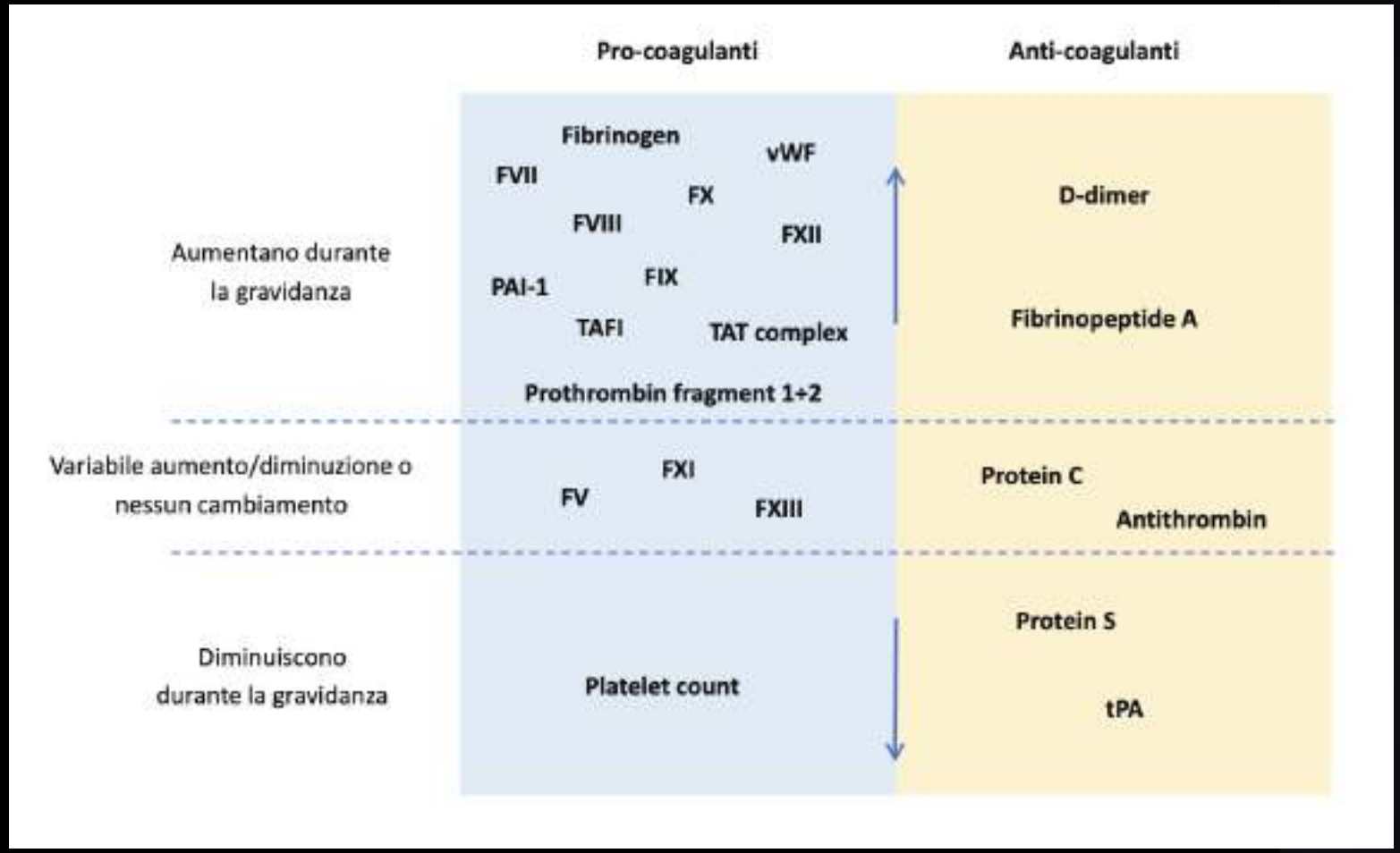
Hypercoagulable state



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- F-VII level increases during pregnancy
- +++ second trimester
- Mild and Moderate FVII deficiency forms
- Levels usually remain insufficient for haemostasis in severely form

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BJOG An International Journal of Obstetrics and Gynaecology

Management of Inherited Bleeding Disorders in Pregnancy

Green-top Guideline No. 71 (joint with UKHCDO)

April 2017

UKHCDO

UNITED KINGDOM HAEMOPHILIA CENTRES DOCTORS' ORGANISATION

Royal College of Obstetricians & Gynaecologists

- Genetic counselling
- Screening
- Risk for baby
- Delivery plan

MULTIDISCIPLINARY TEAM

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GENETIC COUNSELLING/SCREENING

- ✓ Deficit noto
- ✓ Storia clinica di sanguinamento
- ✓ Consulenza Ematologica

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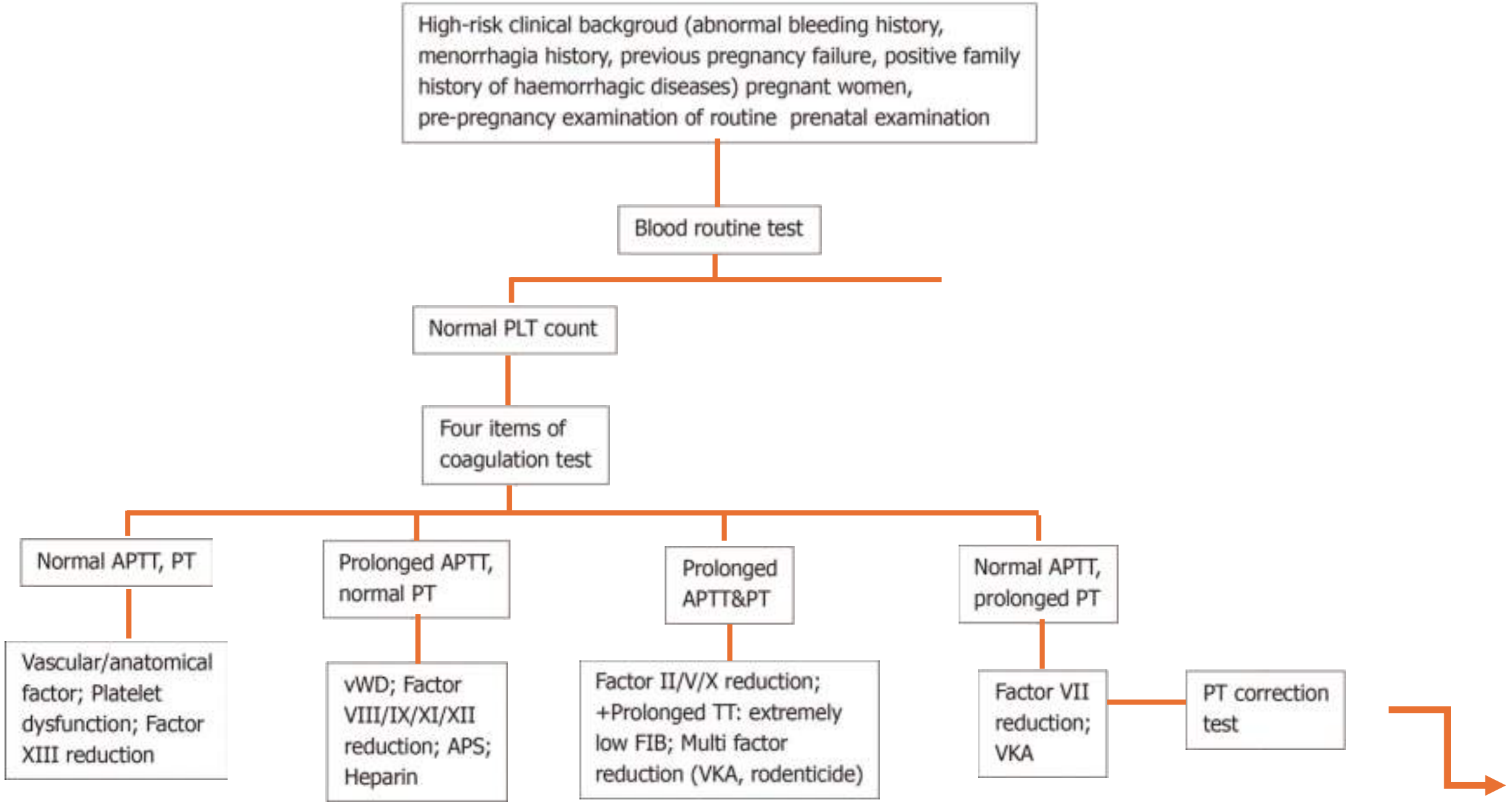
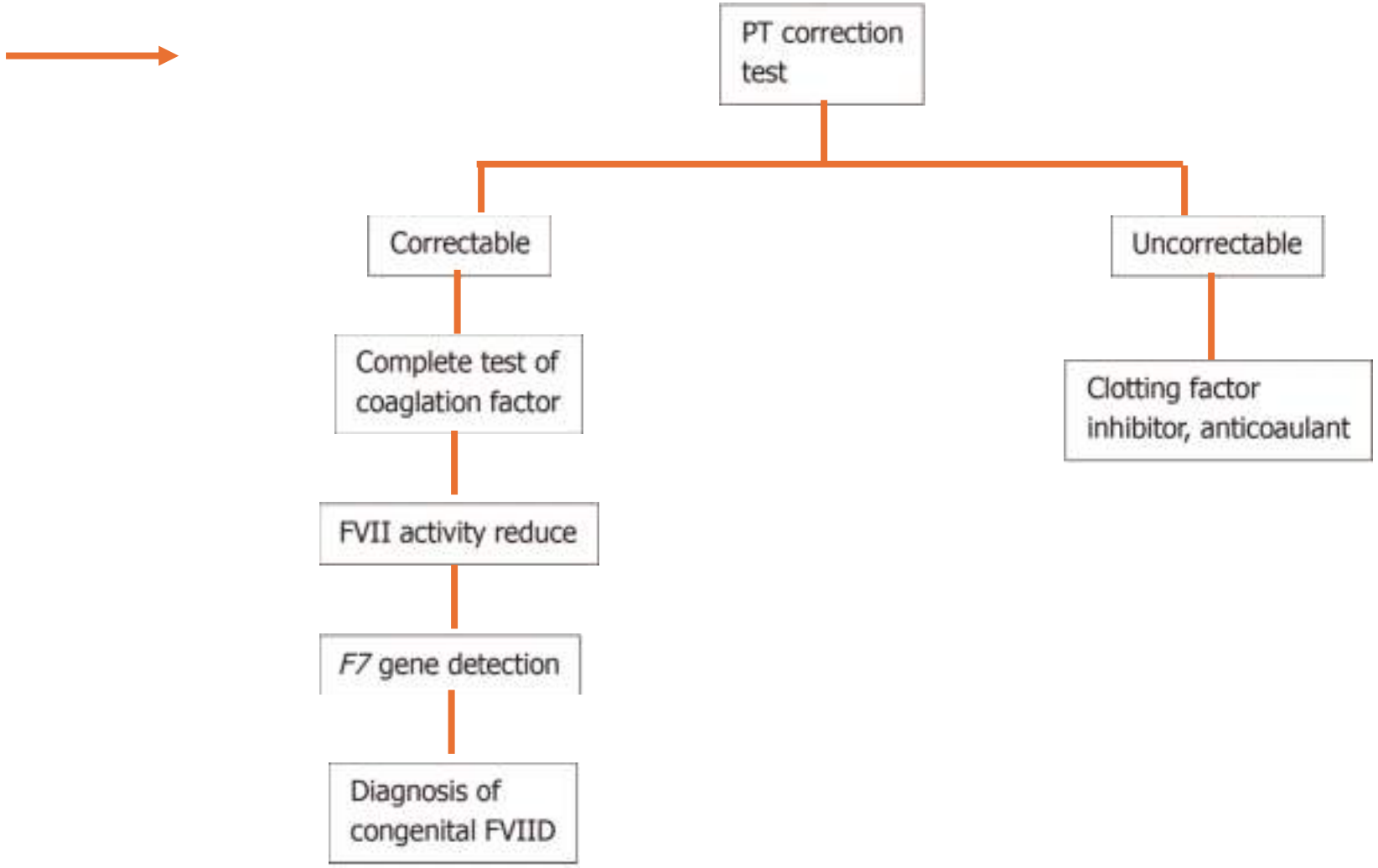
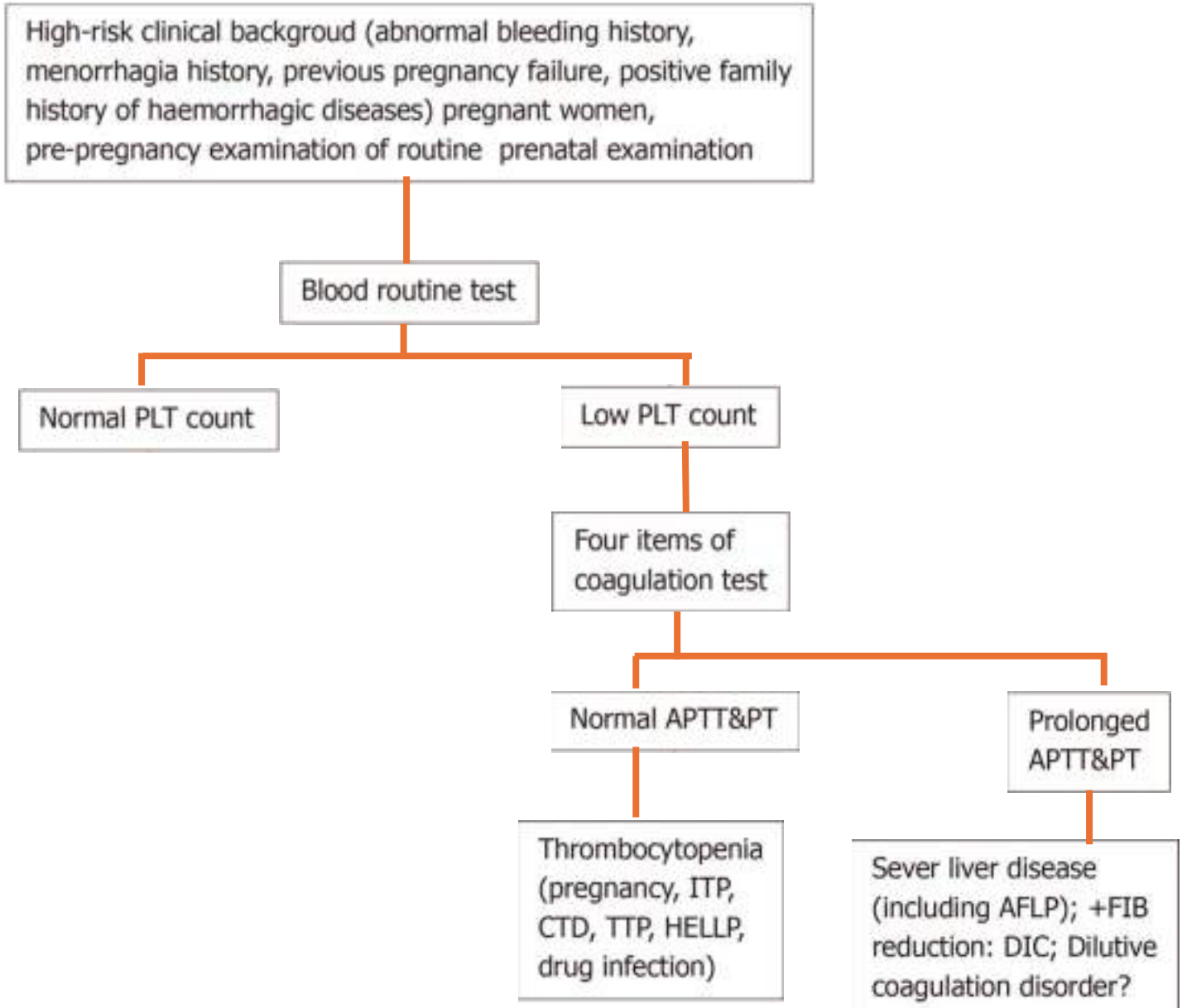
Diagnosis flow chart of pregnancy with congenital factor VII deficiency



Diagnosis and treatment discussion of congenital factor VII deficiency in pregnancy: A case report *World J Clin Cases* 2021



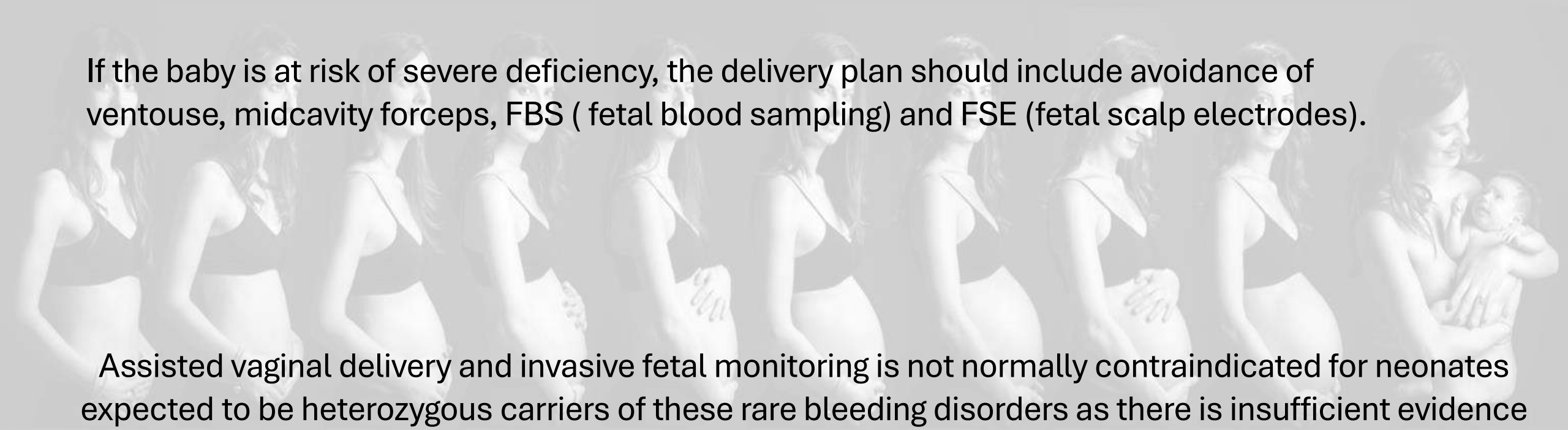
vWD: Von Willebrand disease; APS: Antiphospholipid antibody syndrome; PLT: Platelet; APTT: Activated partial thromboplastin time; PT: Prothrombin time;
TT: Plasma thrombin time; FIB: Fibrinogen; VKA: Vitamin K antagonist; ITP: Idiopathic thrombocytopenic purpura; CTD: Diffuse connective tissue disease;
TTP: Thrombocytopenic purpura; HELLP syndrome; AFLP: Acute fatty liver in pregnancy; DIC: Diffuse intravascular coagulation; FVII: Factor VII; FVIID: Factor VII deficienc



RISK FOR BABY

Babies at risk of homozygosity or compound heterozygosity are at significant risk of bleeding following delivery, including ICH.

If the baby is at risk of severe deficiency, the delivery plan should include avoidance of ventouse, midcavity forceps, FBS (fetal blood sampling) and FSE (fetal scalp electrodes).



Assisted vaginal delivery and invasive fetal monitoring is not normally contraindicated for neonates expected to be heterozygous carriers of these rare bleeding disorders as there is insufficient evidence from case reports to justify restricting these helpful aids to delivery

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Appendix IV: Risk stratification of bleeding risk for the fetus/neonate

Risk stratification of bleeding risk for the fetus/neonate		
Risk level	**Possible or confirmed fetal diagnosis**	**Suggested management of delivery**
High risk	• Males with severe and moderate haemophilia A or B • Type 3 VWD • Severe (homozygous) rare coagulopathies or severe platelet disorders	Consider third trimester amniocentesis to diagnose disease (see PND table). Discuss mode of delivery taking maternal and fetal factors into consideration, but avoid midcavity forceps, ventouse delivery, FBS, FSE, rotational forceps and external cephalic version. Cord sample for factor assay. Oral vitamin K, unless the result is known to be normal.
Medium risk	• Males with mild haemophilia A or B • Type 2 VWD	Consider third trimester amniocentesis to diagnose disease. Avoid midcavity forceps, ventouse, rotational forceps and external cephalic version. Judicious use of FBS and FSE to facilitate vaginal delivery Cord sample for factor assay. Oral vitamin K, unless the result is known to be normal.
Mild risk	• Clinically moderate or severe type 1 VWD in family • Female fetuses who are obligate/possible carriers of severe haemophilia B • Mild platelet function disorders	Consider avoidance of ventouse and external cephalic version. Judicious use of rotational forceps, FBS and FSE.
Unlikely to be at risk	• Clinically mild type 1 VWD in family • All other obligate/possible haemophilia carrier female fetuses • Heterozygous rare coagulopathies	No special precautions.

Abbreviations: **FBS** fetal blood sampling; **FSE** fetal scalp electrode; **PND** prenatal diagnosis; **VWD** von Willebrand disease.

Appendix V: Neonatal bleeding risks and management

Factor	Risk of early bleeding in severe deficiency	Factor/platelet activity in normal term neonates	Diagnosis of severe and moderate deficiency	Diagnosis of mild deficiency[a]
Factor VIII	ICH incidence 1–4%	Normal or mildly increased	Yes	Possible at birth in most cases but re-test at 3–6 months to confirm levels.
Factor IX	ICH reported	Reduced	Yes	Testing required at 3–6 months of age.
VWF	ICH reported	Increased	Yes in type 3 and some type 2 deficiencies	Testing should not be undertaken unless clinically indicated until 6 months of age.
Fibrinogen	ICH and umbilical bleeding reported	Normal or slightly reduced (assay dependent)	Yes	Usually possible at birth. Confirmation required at 3–6 months of age.
Factor II	ICH and umbilical bleeding reported	Reduced	Yes	Testing required at 3–6 months of age.
Factor V	ICH reported	Normal or slightly reduced	Yes	Usually possible at birth. Confirmation required at 3–6 months of age.
Factor VII	ICH reported	Reduced	Yes	Testing required at 3–6 months of age.
Factor X	ICH and umbilical bleeding reported	Reduced	Yes	Testing required at 3–6 months of age.
Factor XI	Spontaneous bleeding seems uncommon Post-surgical bleeding reported	Reduced	Yes	Testing required at 3–6 months of age.
Factor XIII	ICH and umbilical bleeding reported	Reduced	Yes	Testing required at 3–6 months of age. Mild deficiencies may not be clinically significant.
Glanzmann's thrombasthenia	Severe bleeding appears relatively uncommon	Neonatal platelets are generally hyporeactive	Yes. PFA-100 can be used for screening. Definitive testing is PMG analysis by flow cytometry.	NA
Bernard Soulier syndrome	Severe bleeding appears uncommon	Neonatal platelets are generally hyporeactive	Yes. PFA-100 can be used for screening. Definitive testing is PMG analysis by flow cytometry.	NA

TREATMENT OPTIONS

- **<20% IU/dL al 3° trimestre e/o storia di sanguinamento**

QUANDO: A dilatazione Completa nel PS
30-60 minuti prima del TC

DOSAGGIO : rFVIIa 15-30µg/kg ogni 4-6h

DURATA: 3gg post PS
5gg post TC

- **>20% IU/dL al 3° trimestre**

Solo in caso di sanguinamento anomalo: rFVII o Antifibrinolitici

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TREATMENT OPTIONS

rFVIIa: per la sua farmacodinamica attiva l'emostasi nel sito specifico dell'emorragia, per l'interazione con TF.
Per questo motivo è associato ad un rischio trombogenico molto basso (<0.4%)

Acido Tranexamico: 15-30mg/kg o 1gr/die viene utilizzato nelle forme lievi di sanguinamento o in associazione al rFVIIa. Non c'è aumento di rischio trombotico

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- Central Neuraxial Anaesthesia
- Postpartum Pharmacological Thromboprophylaxis
- NSAIDs

USUALLY BE AVOIDED

They may be used after individual assessment if adequate replacement therapy is confirmed.

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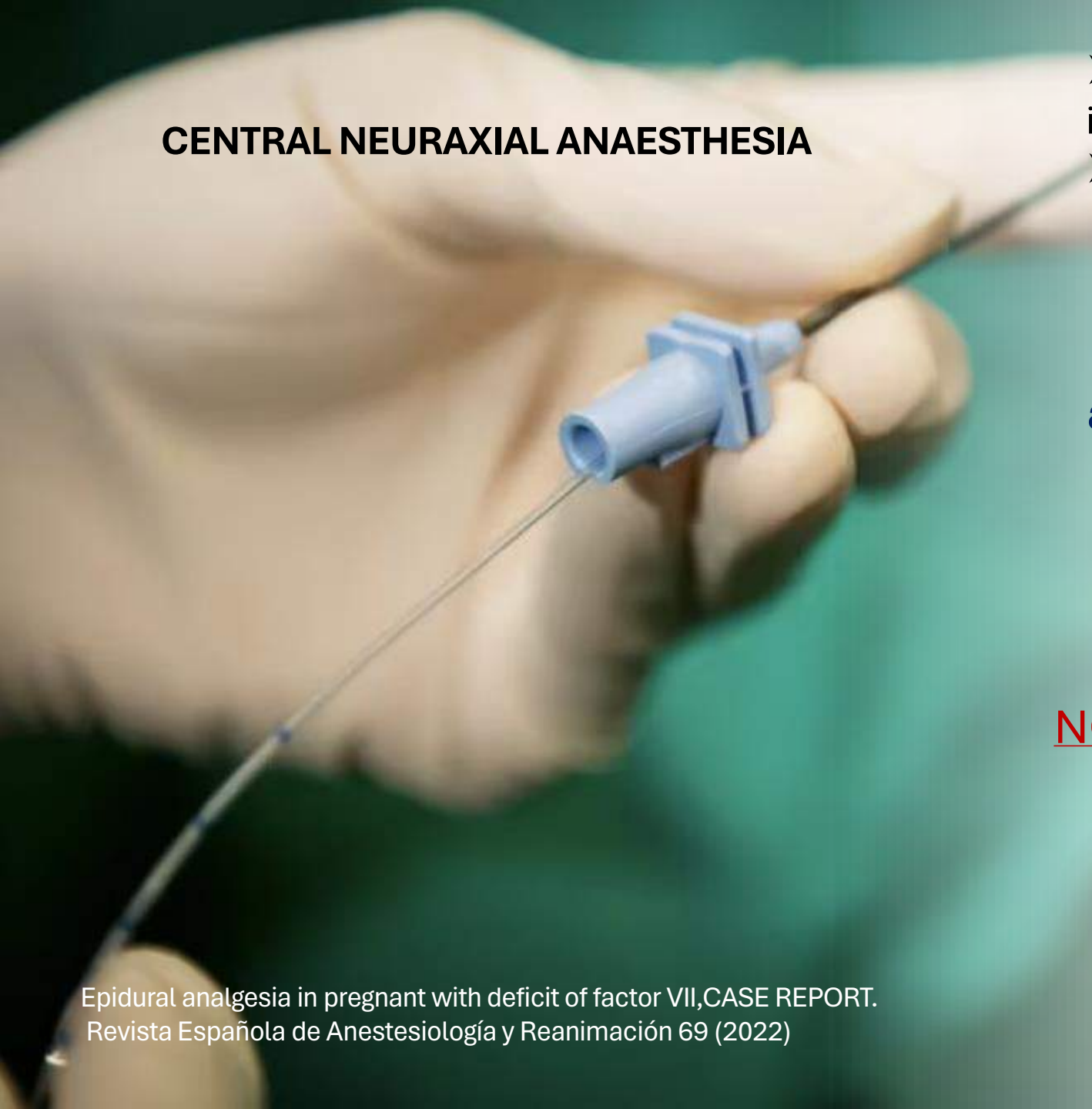
CENTRAL NEURAXIAL ANAESTHESIA



- Scarsità di studi sulle conseguenze dell'ALR in donne con disturbi ereditari dell'emostasi
- Assenza di Linee Guida

«the activity level does not always correlate with bleeding risk, and patients with similar activity levels have variable bleeding tendencies»

Livelli in range di normalità o che rimangono in range dopo rFVIIa:
NON CONTROINDICAZIONE ASSOLUTA ALL'ALR

PROFILASSI: 30-60 min prima di eseguire la procedura

Epidural analgesia in pregnant with deficit of factor VII,CASE REPORT. Revista Española de Anestesiología y Reanimación 69 (2022)

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RISCHIO DI EMATOMA SPINALE

- **Evento raro: 1/150.000-275.000 di APD**

Acta anaesthesiologica Scandinavica, 2016

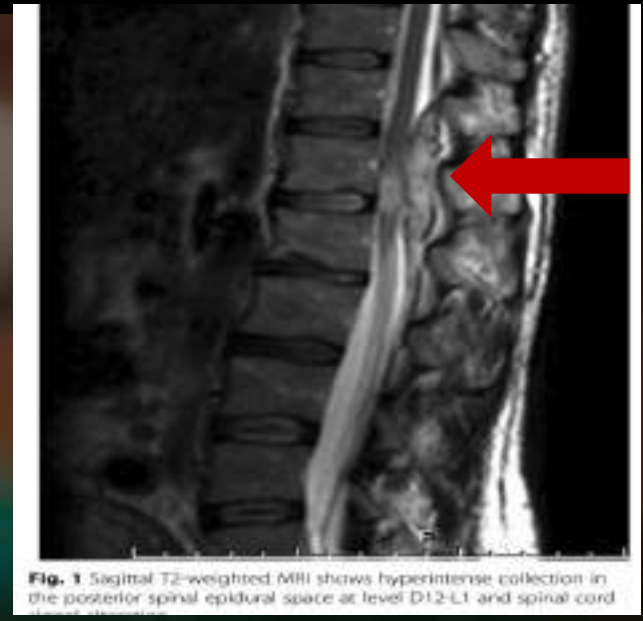
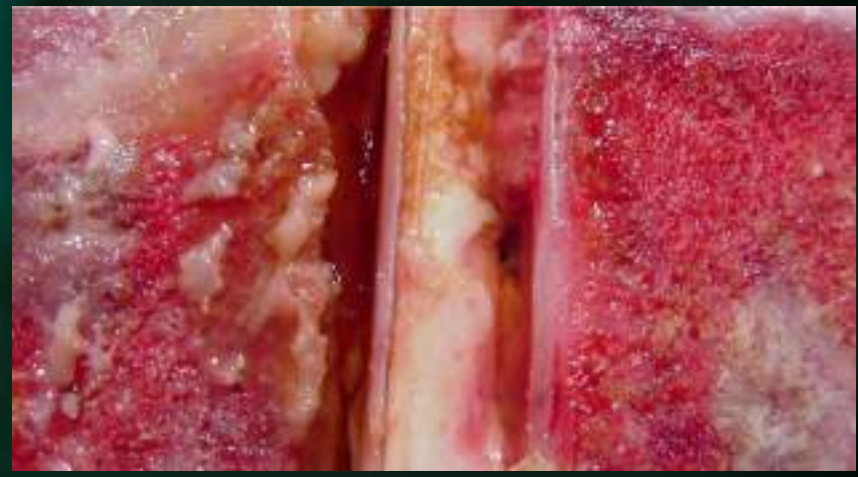


Fig. 1 Sagittal T2-weighted MRI shows hyperintense collection in the posterior spinal epidural space at level D12-L1 and spinal cord

- **Il rischio di ematoma neurassiale nella popolazione ostetrica è stato stimato da Ruppen et al. in 1:168.000**

Anesthesiology 2006; 105: 394-9



- **1: 220 000 - 250 000 in seguito ad AS**

Anesthesiology 2007 105: 394-9

- **Review 2017: 0 eventi con LMWH o UFH profilattica**

Anesth Analg. 2017;125(1):223-231

- **Il rischio di ematoma epidurale e PLT:** **0 - 49.000 11%**

50.000 - 69.000 3% **70.000-100.000 0,2%.**

Anesthesiology, 2017 *126*(6), 1053–1063

Donna di 37 aa, IV G, PARA 0 (3 AS a 4 settimane)
In anamnesi: DEFICIT EREDITARIO del FVII in forma severa (valori basali <10%) e LAC+

APR: metrorragie importanti ni primi mesi del menarca, poi risoltesi. Nessun intervento chirurgico.

Gravidanza insorta con PMA, decorso regolare:
ha assunto CardioASA fino a 32° settimane, poi EBPM

30/05 ricovero per induzione a 38 settimane +4
(ultima assunzione EBPM il 29/05), INR 1.54

RISCHIO EMORRAGICO MEDIO-ELEVATO

PROTOCOLLO DI GESTIONE

condiviso tra ginecologi, ostetriche, anestesisti, ematologo, neonatolo

- Somministrare FVII 15mcg/kg PRIMA dell'analgesia neuroassiale
- AL momento del parto: Ac tranexamico 1 gr
- Se evidenza di EPP: FVII 90mcg/kg

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01/06 posizionato CRB , poi rimosso e
proseguita induzione con MISOPROSTOLO

Dopo VI dose di MISOPROSTOLO,
intensa attività contrattile,
Trasferimento in Sala Parto

Ore 01:50 somministrato rFVIIa 15mcg/kg

Ore 03:40: collo centralizzato , appianato,
DC 3 cm, PP cefalica -2, MAC rotte, LC.
Presenza di attività contrattile dolorosa:
si richiede PARTO ANALGESIA, INR 0.72

Travaglio regolare, continua
induzione con ossitocina

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Ore 19:58 PARTO OPERATIVO per esaurimento delle forze materne, secondamento spontaneo.

Al momento del parto somministrare:
Ossitocina X UI
Sulprostone 500mcg ic
Acido Tranexamico 1gr
FVIIa 15mcg/kg

PERDITE EMATICHE STIMATE: 40Oml
INR post partum: 0.72,
controllo successivo INR: 0.97
Rimosso Catetere peridurale.
Iniziata profilassi con EBPM
In III GPO: Dimissione, INR 1.39

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La frase più pericolosa in assoluto è:

"Abbiamo sempre fatto così".

Grace Murray Hopper

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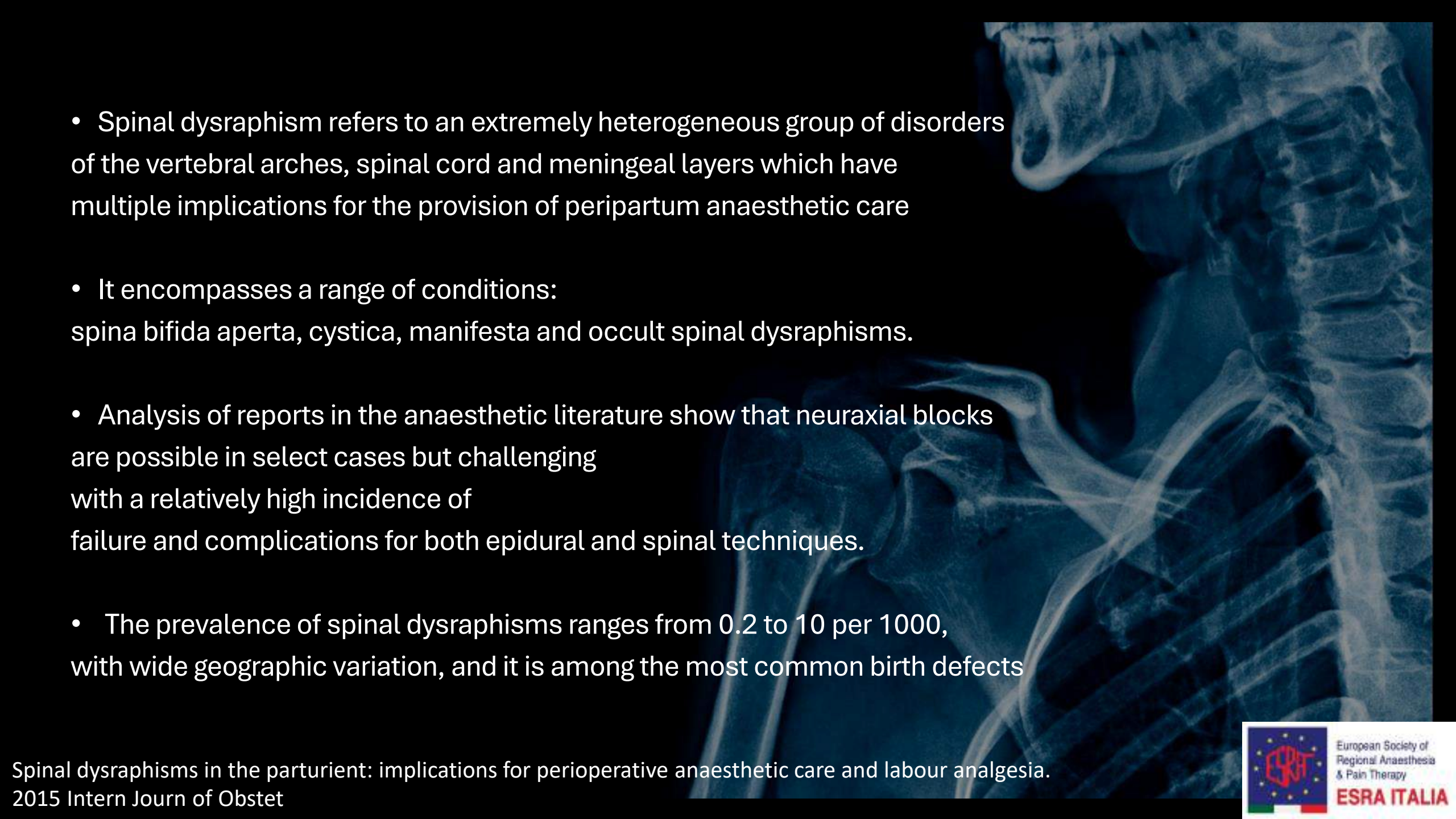

- Spinal dysraphism refers to an extremely heterogeneous group of disorders of the vertebral arches, spinal cord and meningeal layers which have multiple implications for the provision of peripartum anaesthetic care

- It encompasses a range of conditions:
spina bifida aperta, cystica, manifesta and occult spinal dysraphisms.

- Analysis of reports in the anaesthetic literature show that neuraxial blocks are possible in select cases but challenging
with a relatively high incidence of
failure and complications for both epidural and spinal techniques.

- The prevalence of spinal dysraphisms ranges from 0.2 to 10 per 1000,
with wide geographic variation, and it is among the most common birth defects

Spinal dysraphisms in the parturient: implications for perioperative anaesthetic care and labour analgesia.
2015 Intern Journ of Obstet

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Classification Tortoni-Donati

uses a combination of clinical and radiological assessment

Table 1 Classification of spinal dysraphisms[5]

Open spinal dysraphisms	
Myelomeningocele	
Meningocele	
Hemimyelomeningocele	
Hemimyelocele	
Closed spinal dysraphisms	
With subcutaneous mass	Without subcutaneous mass
Lumbosacral	*Simple dysraphic states*
Lipomyelocele	Posterior spina bifida
Lipomyelomeningocele	Lipoma
Meningocele	• Intradural
Terminal myelocystocele	• Intramedullary
	• Filum terminale
Cervical	Tight filum terminale
Meningocele	Abnormally long spinal cord
Myelocystocele	Persistent terminal ventricle
Myelocele	
	Complex dysraphic states
	Dorsal enteric fistula
	Neurenteric cysts
	Split cord malformations
	• Diastematomyelia
	• Diplomyelia
	Dermal sinus
	Caudal regression syndrome
	Segmental spinal dysgenesis

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- Neurological impairment, manifest as motor and sensory dysfunction, absent reflexes, sphincter dysfunction, hydrocephalus and Chiari II malformations were more common with higher lesions and those that were classified as “open’’ at birth
- The majority of these abnormalities are vertebral arch defects in the sacrum, with 80% occurring at S1, 10% at S1–2, 8.4% at L5 and 0.2% at L5–S1.

- The clinical significance of such findings in asymptomatic patients is disputed and is largely considered to be a variant of normal.

Table 2 Clinical manifestations of spinal dysraphisms

Cutaneous	Urological[60]	Neuro-orthopedic[60]
High index of suspicion	Incontinence	Talipes equinovarus
Hypertrichosis	Recurrent UTI	Pes cavus
Dimples		High arches
• Large		Hammer toes
• >2.5 cm from anal margin		Clawed feet
Acrochordons		**Asymmetry**
Pseudo-tail		Buttock
True tail		Leg
Haemangiomas		Foot
Aplasia cutis/scar		**Symptoms**
Dermoid sinus or cyst		Non dermatomal back pain
		Numbness
Low index of suspicion		Weakness
Telangiectasia		
Capillary malformation (port wine stain)		
Hyperpigmentation		
Melanocytic nevi		
Teratomas		

UTI: urinary tract infection.

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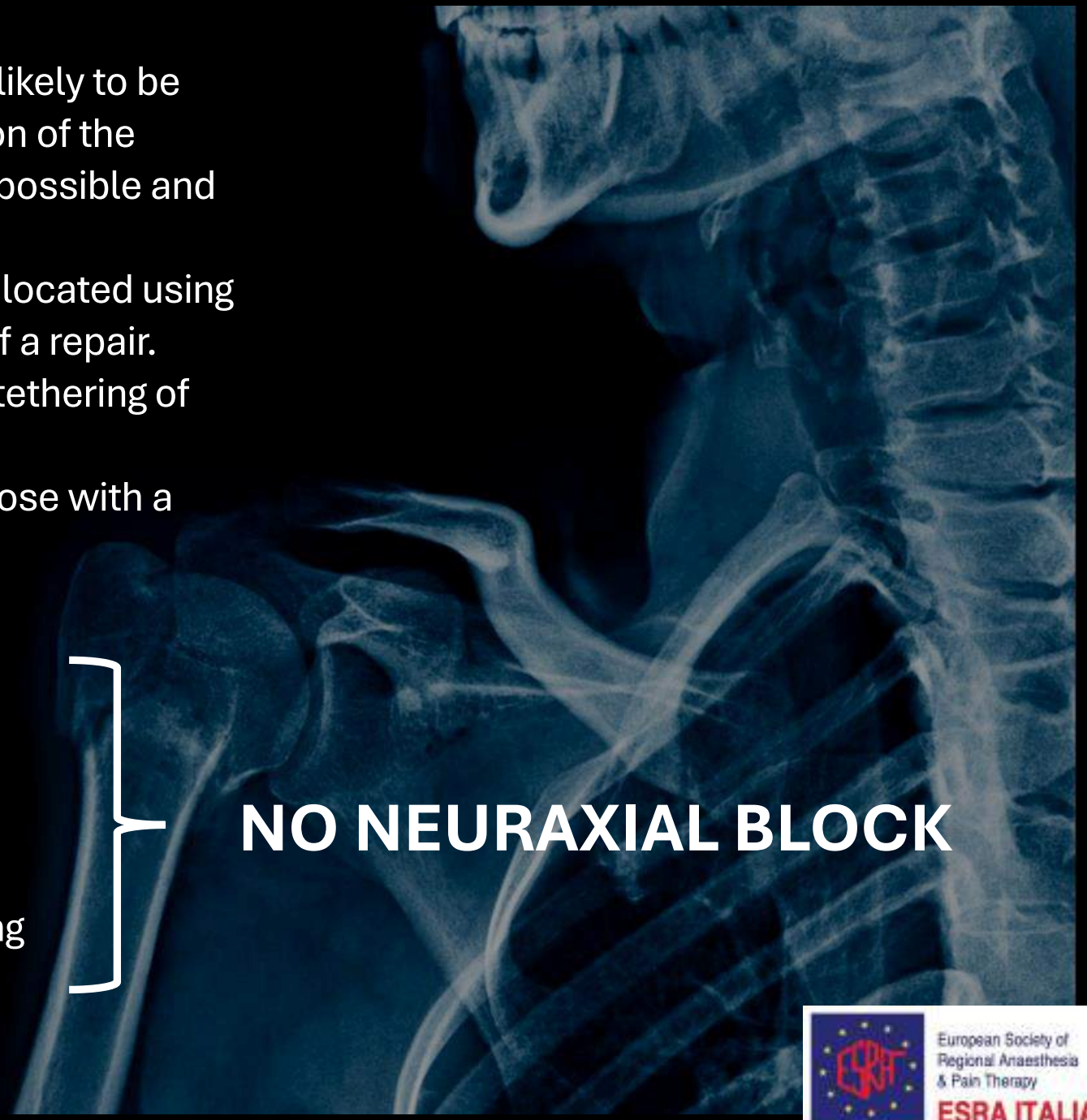
After surgical repair, the epidural space is unlikely to be normal and can be non-existent. Identification of the vertebral level at the site of the defect is not possible and the ligamentum flavum is not present.
Consequently, the epidural space cannot be located using a loss-of- resistance technique at the level of a repair.
In patients who have undergone surgical de-tethering of the spinal cord, re-tethering (which may be asymptomatic) is common, particularly in those with a posterior dural attachment before surgery

- ✓ History of spinal fusion
- ✓ Tethered cord
- ✓ Previous dural puncture
- ✓ Existing neurological deficit
- ✓ Absence of appropriate diagnostic imaging

NO NEURAXIAL BLOCK



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Neuraxial analgesia for labour or C-Section ?????

ALR in casi selezionati

Valutazione clinica e Neurologica

Operatore esperto

Eseguire la procedura a livelli anatomicamente normali con legamento giallo intatto.

Evitarla in zone con cicatrici e/o lesioni

Possibile analgesia/anestesia incompleta su pregressi interventi chirurgici

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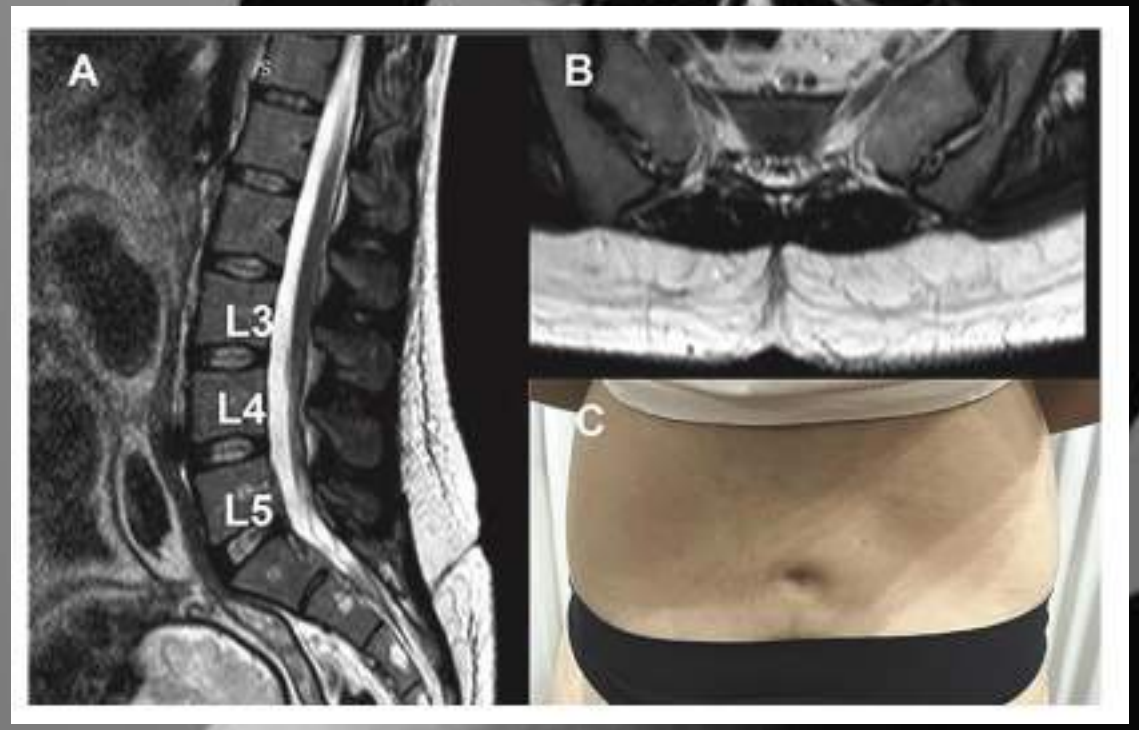
Donna 30 anni, ASA 2

38 settimane

EO: fossetta cutanea livello sacrale.
No scoliosi visibile

EON: assenza di deficit sensitivo motori

RMN lombare: midollo ancorato con evidenza di
interruzione arco vertebrale posteriore a livello
di S2 e associato cono midollare posizionato in modo anomalo



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In travaglio:

Analgesia con Tecnica EPIDURALE
Livello L2-L3
Farmaci: Sufentanil 10mcg+
Top up con Ropivacaina 0.1-0.15%

Gestione ottimale del dolore
Non complicanze anestesiologiche né ostetriche
Non complicanze neurologiche

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“ Consegnare il bambino tra le braccia di una mamma cosciente e senza dolore è uno dei momenti più eccitanti e gratificanti in medicina”

Donald Moir
Founder President Of The Obstetric
Anesthetist’s Association

Grazie