

## Venous access in children undergoing ITI

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### ITI

- ITI – different regiments
  - factor FVIII/FIX concentrates and/or by-passing agents administered i.v.
  - frequency from 3 x week to 2 x day
- sufficient and convenient venous access necessary!




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### I.v. routes

- peripheral vein
- central vein
  - central venous access devices
    - short time CVL
    - long time CVL
      - tunnelled
      - port-a-cath
- A-V fistula




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### Peripheral veins

- painful venepuncture
- need for good compliance ("stillness")
- local anaesthetics can help
  - EMLA crm 5%
    - (lidocain+prilocain) 1-2 hours prior puncture in occlusion



### Peripheral veins

Gauge	24	22	20	18
mm	0,7	0,9	1,1	1,3

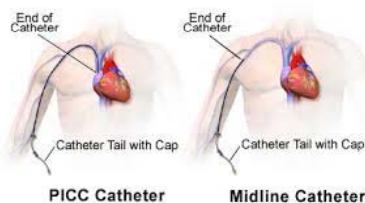


Vasofix inserted in a vein up to 72 hours



### Peripherally Inserted Central Catheters

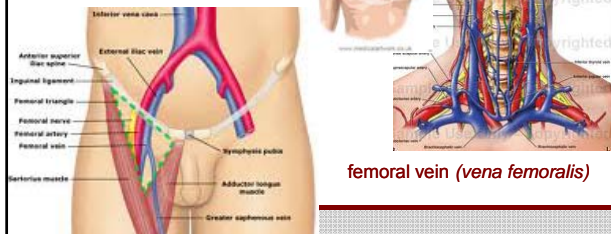
- higher risk of thrombosis
- lower risk of occlusion
- no data from haemophilia patients



## Central Venous Access Devices

jugular vein (*vena jugularis*)

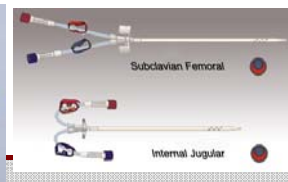
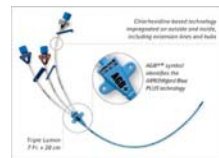
subclavian vein (*vena subclavia*)



femoral vein (*vena femoralis*)

## Central Venous Access Devices

- external short term use
  - up to 4 weeks
  - emergency situations
  - after surgery




## Central Venous Access Devices

- external long-term use
  - tunneled
    - Hickmann, Broviac
  - risk of infection
  - risk of self extraction
  - sterile techniques
  - regular flushing
    - saline, heparin



### Central Venous Access Devices

- internal
  - ports (port-a-cath)




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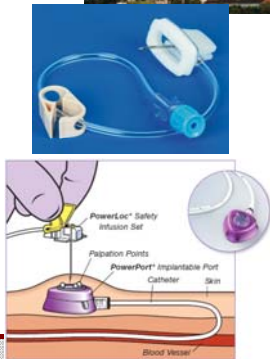
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### Central Venous Access Devices - Ports

- lower risk of infection
- longer life
- no risk of “self pulling”
- bathing possible
- **puncture with needle required**




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### CVAD infections

- exit site infection
- tunnel or pocket infection
- sepsis




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### CVADs: infection



- pooled incidence of infection 0.66/1000 catheter days
- factors associated with an increased risk of infection
  - presence of inhibitors
  - external lines
  - age 2-6 years
  - daily use
- reason for removal
  - 70% infection
  - 4% thrombosis

(Valentino et al, Haemophilia 2004)

- CVAD infection = lower ITI success  
(I-TI study)

- 81 CVADs, 3.36 years (0.22 - 9.44 years), Median age at first CVAD insertion 2.16 years (0.7- 14)
- Overall incidence of confirmed CVAD-related bloodstream infection 0.42 per 1000 CVAD days, incidence of confirmed and suspected CVAD-related infection 0.60 per 1000 CVAD days
- 73.7% successfully treated iv ATB without CVAD removal  
(Yeoh et al, J Paediatr Child Health. 2013)

### CVADs: thrombosis



- even in haemophilia patients
- asymptomatic
  - on CVL
  - mural
  - complete occlusion
  - risk of progression (VSC+VJI+VBC...)

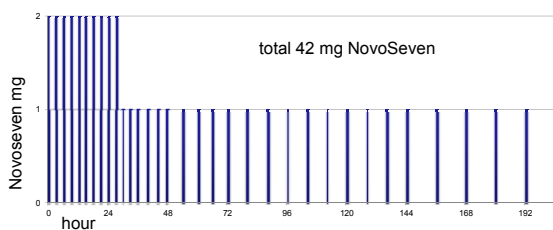
- locus of infection  
(Santagostino et al Blood Transfus 2008)

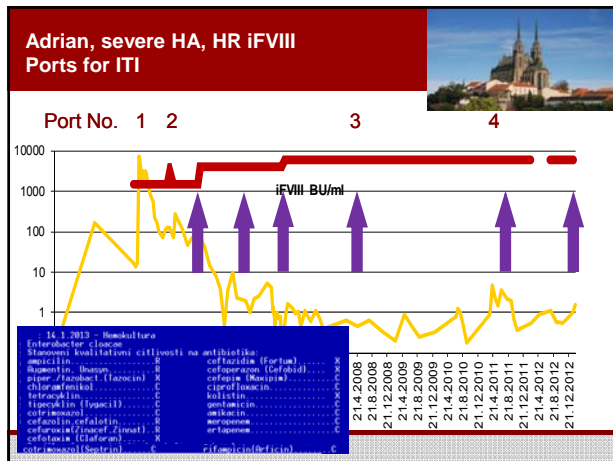
- 20 boys with haemophilia with total 27 CVADs
- MRI diagnosed DVT in 5 (25%), clinically silent, all inserted below 1 year
- no correlation between the duration or number of CVADs and DVT was detected. None of the patients had subjective symptoms of PTS  
(Ranta et al, Haemophilia 2012)

### Management of port insertion Kryštof, severe HA, HR iFVIII 35 BU/ml, 18 months



- Port-a-cath insertion for ITI, 12 kg
- rFVIIa NovoSeven 1 mg = 83 ug/kg 2 mg = 166 ug/kg
- + paraaminobenzoic acid





**AV fistulas**

- alternative to CVADs
- in haemophilia patients since 1999  
(Santagostino et al BJH 2003)
- surgeon experience
- non-dominant upper limb preferred

**AV fistulas**

- data from Italy, USA, Austria
- 1 month to maturation
  - dilatation and arterialisation of the vein
- 82% successful maturation
- duration of use 1-7 years (median 5 years)

(Santagostino et al, BJH 2003, JTH 2007  
McCarthy et al J Vasc Surg 2007)

## AV fistulas



- complications
  - hematoma
  - distal limb ischemia
  - thrombosis of venous part
  - loss of patency
  - limb hypertrophy
  - aneurysmatic dilatation



## Summary



Type	+	-
PV	no external device no surgery, no special care	not sufficient in small boys haematoma post puncture
External CVL	no needle immediate use percutaneous insertion	infection, thrombosis limited duration (year) self-displacement sterile techniques
Internal CVL	longer duration (years) no physical activity limits	surgical insertion infection, thrombosis needles, skin erosion sterile techniques
AVF	no infection, no special care longer duration (years) no physical activity limits	surgical construction delayed use AVF complications

## Conclusions



- adequate venous access is one of basic condition for successful ITI
- peripheral vein is a best option
- CVAD or AV fistulas are possibilities
- individual approach to patient



Future

- long acting drugs
- s.c. formulations
- p.o. formulations



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Thank you for your attention



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