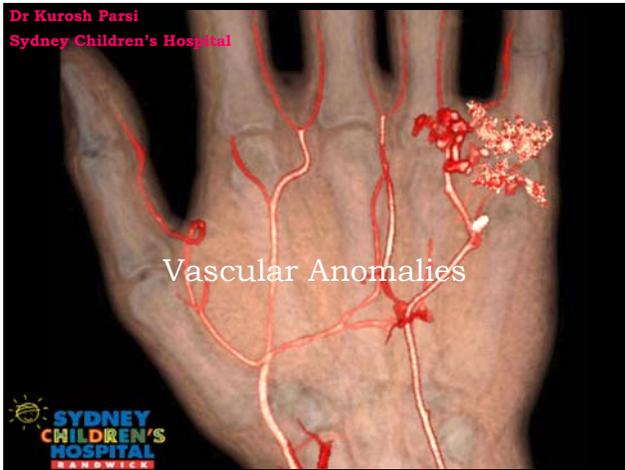


Dr Kurosh Parsi  
Sydney Children's Hospital



## Vascular Anomalies



## ISSVA Classification

International Society for the Study of Vascular Anomalies



Tumours	Malformations	
	Simple	Combined
Haemangioma	Capillary (C)	Arteriovenous fistula (AVF)
Other Tumours	Lymphatic (L)	AVM
	Venous (V)	CVM
		CLVM
		LVM
		CAVM
		CLAVM

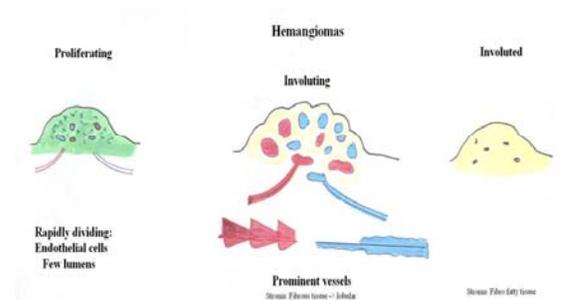
Scientific Committee of the Eleventh Meeting of the International Society for the Study of Vascular Anomalies (ISSVA), Rome, Italy, 1998

## Haemangioma of Infancy

### ▶ Natural history:

- ▶ Not present at birth
- ▶ More prominent days to weeks after birth
  - ▶ Proliferate over ~6 months
  - ▶ Most growth complete by 4mo
- ▶ Involution at 9-12 months
- ▶ Residual fibrofatty tissue after involution
- ▶ Can cause significant complications

## Hemangioma of Infancy





## Vascular Malformation

- ▶ Present at birth
- ▶ Usually grow in proportion to child
- ▶ Never regress



## Capillary Malformation



## Venous Malformations

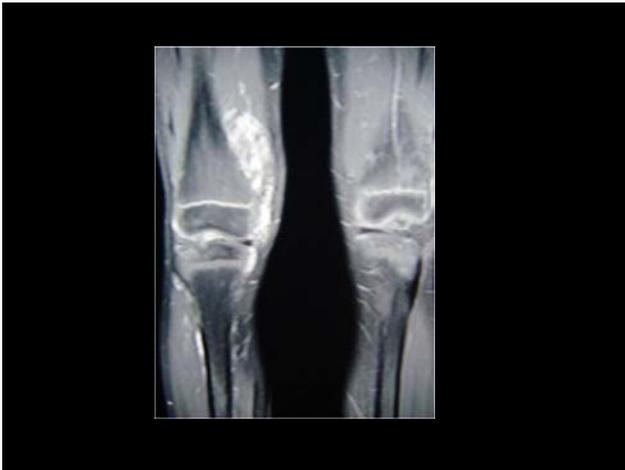
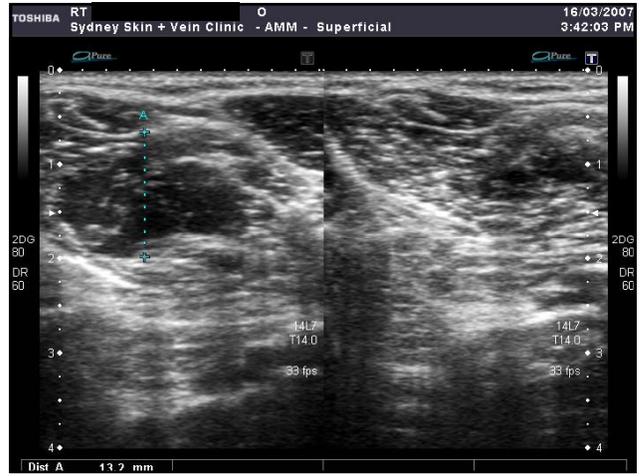
## Venous Malformations

- ▶ **Truncular**
  - ▶ IVC Anomalies
  - ▶ Primary venous aneurysms
- ▶ **Extra-truncular**
  - ▶ Intra-dermal
  - ▶ SC fat
  - ▶ Intra-muscular
  - ▶ Intra-articular
  - ▶ Deep within other organs

## Complications

- ▶ **STP → DVT → PE**
  - ▶ When extensive positive d-dimers
- ▶ **Involve important structures**
  - ▶ Airways
  - ▶ Eyes → extension to brain
  - ▶ Perineum
  - ▶ Intra-articular
- ▶ **Chronic venous hypertension → LDS → Ulcers**
- ▶ **Limb hypertrophy**

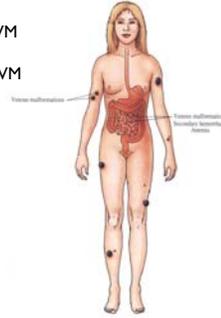




## Phlebectatic Venous Malformations

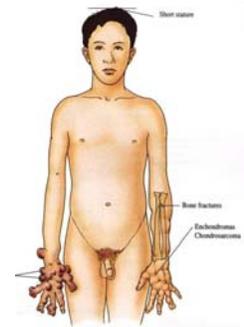
## Blue Rubber Bleb (BRB) Syndrome

- ▶ Inherited AD
- ▶ Multiple VM
- ▶ DD of GVM
- ▶ Multiple cutaneous and GI lesions
- ▶ GI Lesions
  - ▶ Bleed causing Fe def
  - ▶ GI infarction
- ▶ Spontaneous thrombosis
- ▶ Other organs involved:
  - ▶ Brain, heart, lung, nasopharynx



## Maffucci Syndrome

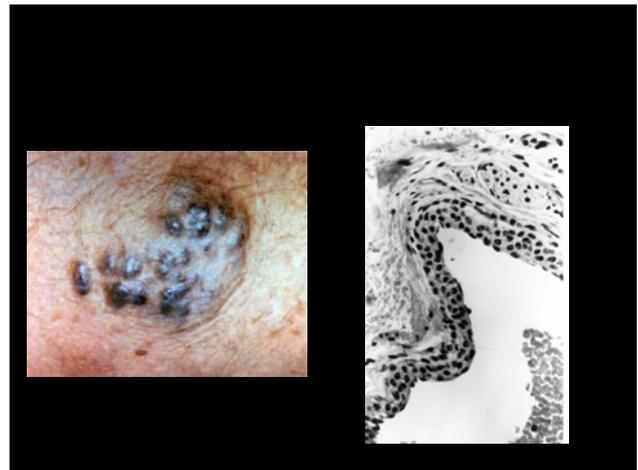
- ▶ VM, AVM
- ▶ Enchondromas
- ▶ Bony abnormalities
  - ▶ Usually asymmetric
  - ▶ Cause secondary fractures





### Glomovenous Malformations

- ▶ Arise from glomus cells (Smooth m. origin)
- ▶ Glomus bodies
  - ▶ AV shunts located acrally
  - ▶ Thermoregulatory receptors



## Pre and Post Sclerotherapy



## Lymphatic Malformations

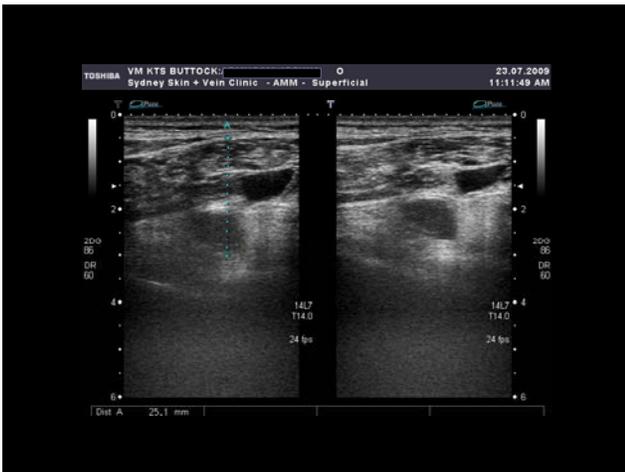
### Lymphatic Malformations

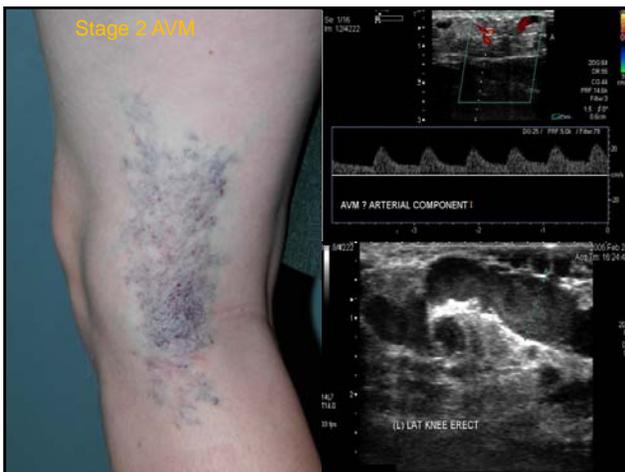
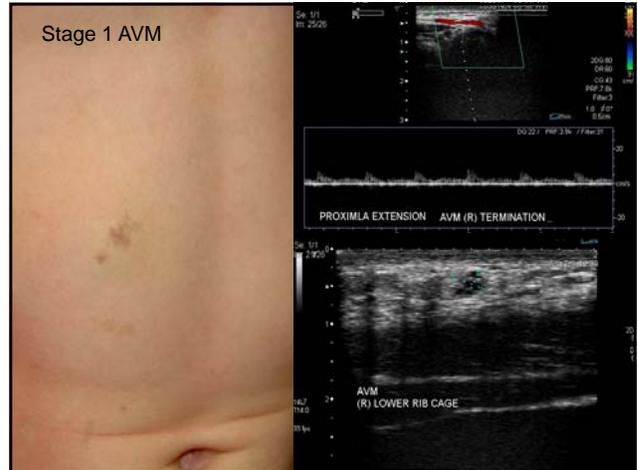
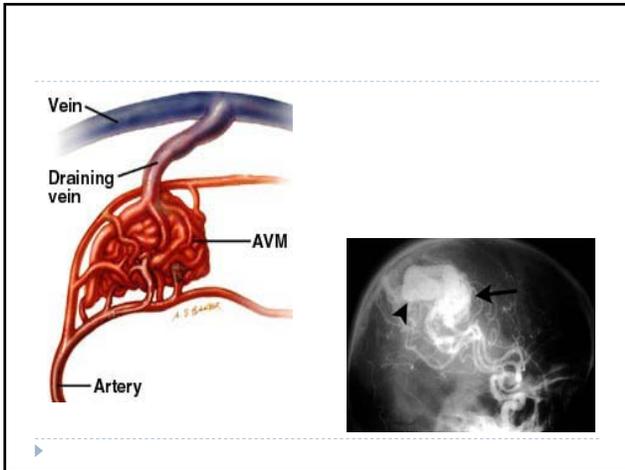
- ▶ **Truncular**
  - ▶ Presents as primary lymphoedema
- ▶ **Extra-truncular**
  - ▶ Macrocystic
  - ▶ Microcystic

### Lymphatic Malformations

- ▶ **Cutaneous complications**
  - ▶ Lymphoedema
  - ▶ Papillomatosis
  - ▶ Infection
  - ▶ Bleeding
  - ▶ Thrombosed lesions







## AVM

### ▶ Chronic Aorto-Caval fistulae

- ▶ Bilateral severe 'varicose veins'
- ▶ Arterial flow in veins
- ▶ 'Machinery' abdominal murmur
- ▶ Abdominal pain, haematuria

### ▶ May lead to

- ▶ Limb/tissue hypertrophy
- ▶ Tachycardia
- ▶ Skin atrophy, ulceration



## Complex Malformations

## Complex Malformations

### ▶ Klippel Trenaunay Syndrome

- ▶ C + V + L

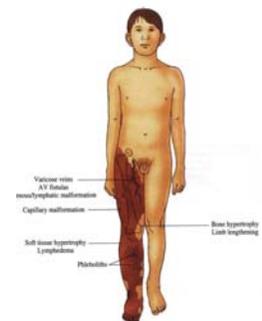
### ▶ Parkes Weber Syndrome

- ▶ C + V + A + L

## KTS

### ▶ Commonly presents as 'varicose veins'

- ▶ Present at birth
- ▶ Usually unilateral
- ▶ Single limb
- ▶ An arm and a leg on the same side





Courtesy of Prof Hugo Partsch

### Parkes Weber Syndrome CLAVM



### QUIZ

- ▶ Guess the malformation from the ultrasound images

## Vascular Malformations

**TIF**

- ▶ Normal structures; abnormal function
- ▶ Error of morphogenesis ( 4<sup>th</sup>-10<sup>th</sup> week)
- ▶ Not present at birth
- ▶ Grow proportionately to the child
- ▶ 90% involute by the age of 9

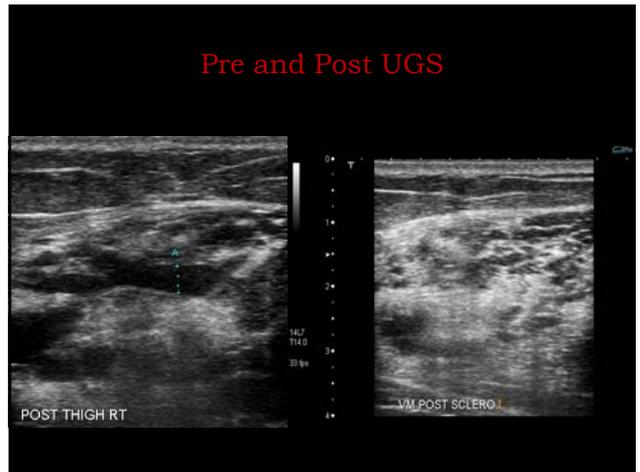
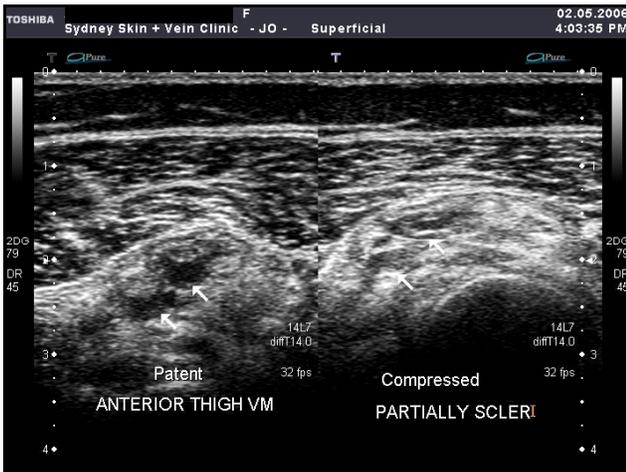
## Doppler Findings

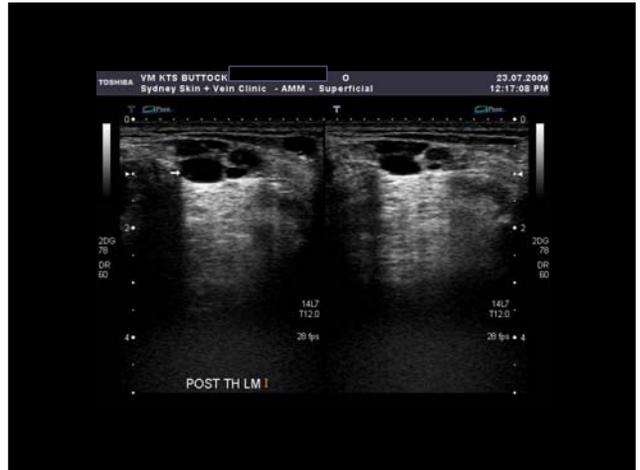
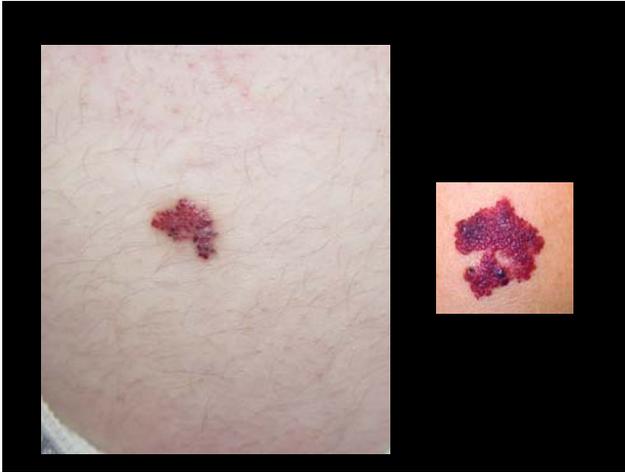
- ▶ High Flow lesions
  - ▶ Haemangiomas
  - ▶ Venous
  - ▶ AVM
  - ▶ Lymphatic
  - ▶ AVF
  - ▶ Capillary

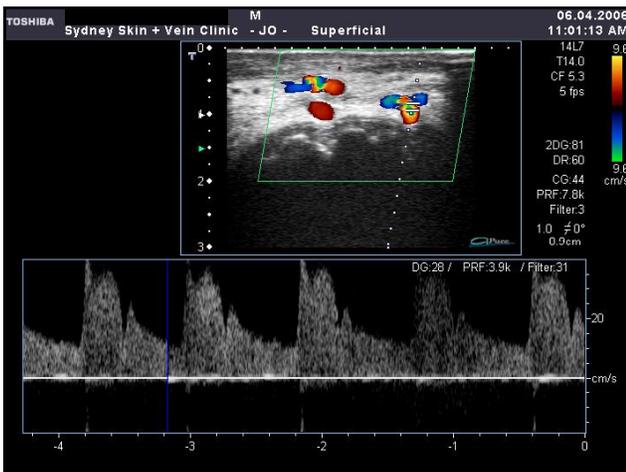
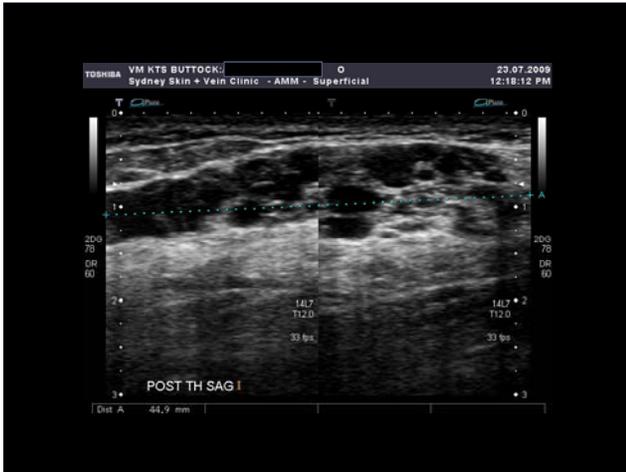
**TIF**

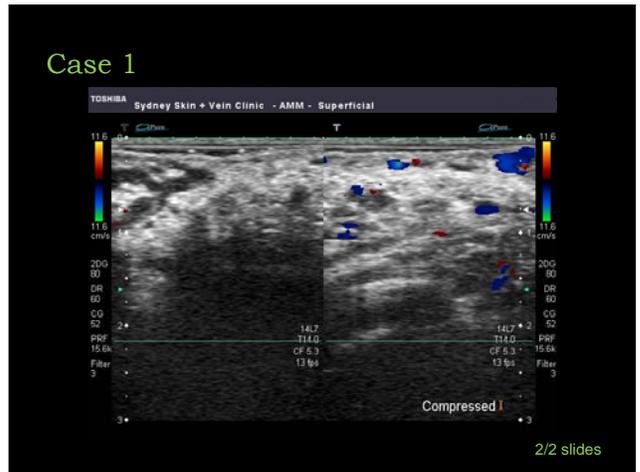
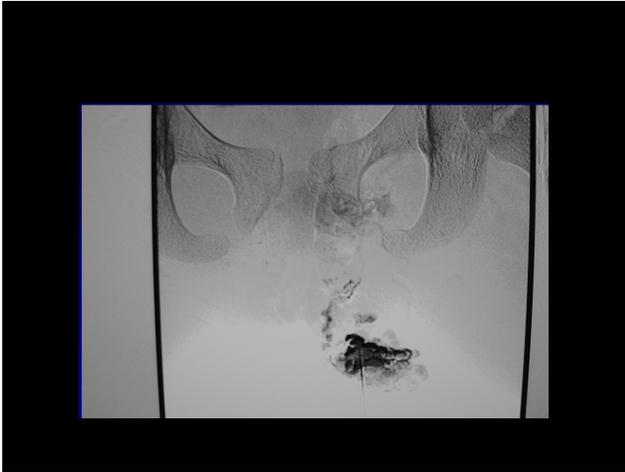




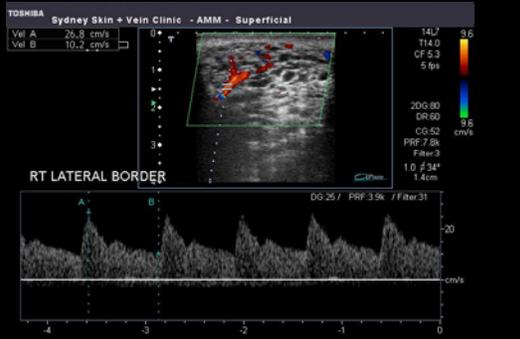




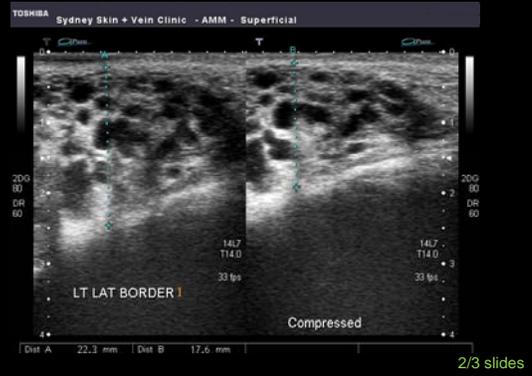




## Case 2



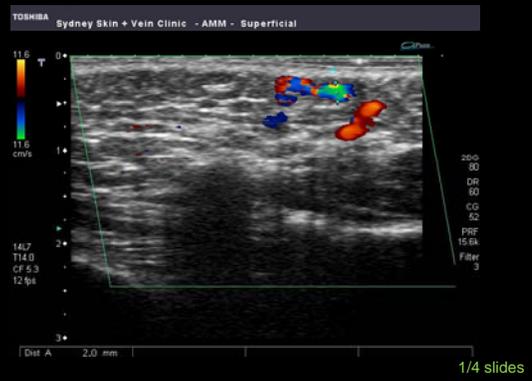
## Case 2



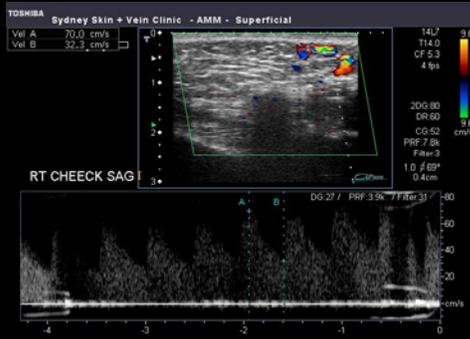
## Case 2



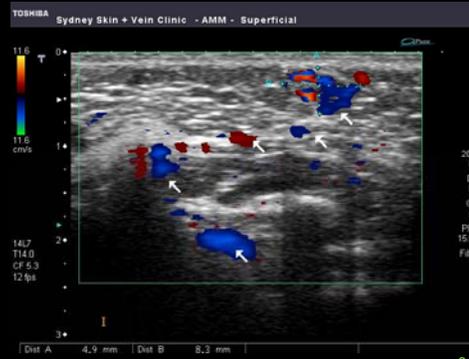
## Case 3



### Case 3



### Case 3



### Case 3



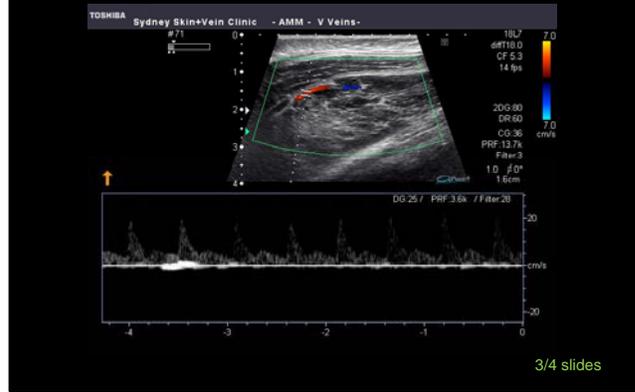
### Case 4



### Case 4



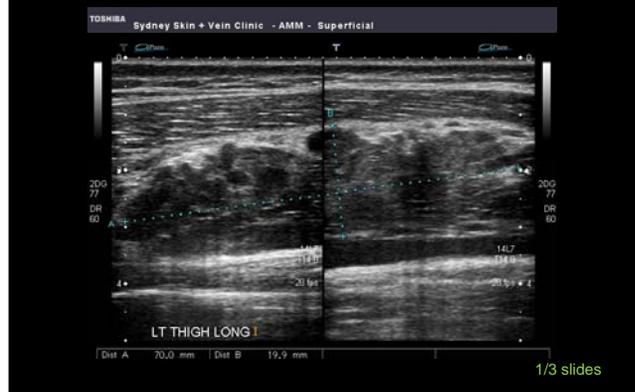
### Case 4



### Case 4



### Case 5



### Case 5



### Case 5

