team. Majority (75%) of the handovers were presented by registrars/junior trainees with only 35% receiving any feedback.

SBAR (Situation, Background, Assessment and Recommendation) method was only used for 42% of handovers. Majority (70%) of the handovers were conducted with the aid of printed sheets, which included: patient demographics (83%), presenting complaints (85%), investigations, results and treatment plans (83%). Only 11% of handovers were done electronically. Handovers had allocated start times (96%) with designated places (89%) close to area of work. However only 65% of the handovers started on time, 20% were free from distractions by allied professionals and just 5% were ‘bleep’ free. 68% had some educational activity within the time allocated in the handover. WPBAs were initiated or completed in only 11% of handovers. Overall 91% of trainees felt that the quality of handover was either average or good.

Conclusions The findings from our survey suggest that the quality of handovers is variable. Handovers should have a structured approach and free from distractions to ensure safety and continuity of care. Incorporating formal teaching and WPBA’s could help develop the role of handovers.

**Results**

1. All rotating registrars have had exposure to pigtail chest-drain insertion in simulation setting and subsequently went on to undertake these skills in NICU on real patients with greater confidence.

2. Improved team working observed between doctors and nursing staff on NICU

**Conclusions** Our method of manikin manipulation is innovative, affordable and effective and can be implemented in any hospital setting to teach practical neonatal skills, improve team working, enhance competency at performing practical skills and work with increased confidence.

### Clinical Genetics Group/British Society of Paediatric Dermatology

### G24 CAPILLARY MALFORMATIONS – ARTERIOVENOUS MALFORMATIONS/ARTERIOVENOUS FISTULA SYNDROME (CM-AVM SYNDROME): AN UNDER RECOGNISED CLINICAL ENTITY?

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**Background** Hereditary Hemorrhagic telangiectasia (HHT) tends to be the first condition to be considered in the differential diagnosis of patients presenting with high flow vascular malformations in combination with cutaneous vascular lesions. However, particularly in the paediatric population, capillary malformation-arterial venous malformation syndrome (CM-AVM) due to RASA-1 mutation is more likely.

**Aims** To present the clinical features of three patients with CM-AVM syndrome, promote knowledge of this condition and aid prompt diagnosis.

**Methods** Clinical examination, detailed family history, imaging (ultrasound, MRI, angiography) and genetic testing.

**Results** Patient 1 was born with a large vascular mass affecting the right side of the face and multiple cutaneous capillary malformations. Patient 2 had a spinal AV fistula and two vascular stains. Patient 3 presented with an intracranial haemorrhage secondary to a parietal AVM and was noted to have several cutaneous vascular lesions. Patients 2 and 3 were referred to the dermatology team as suspected HHT. The cutaneous vascular lesions present in all three patients were consistent with capillary malformations (in keeping with a diagnosis of CM-AVM) and were not typical of telangiectases.

**Conclusion** In patients with high flow CNS vascular lesions, it is crucial to establish the precise nature of cutaneous vascular lesions in order to request appropriate genetic testing and screening of relatives.

**REFERENCE**

1. Laurence M Boon, Nicole Revenu, Mikk a Vikkula, Université catholique de Louvain, Brussels, Belgium.

### G25 RASA1 MUTATIONS AND VEIN OF GALEN ARTERIAL MALFORMATIONS

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**Introduction** Capillary malformations (CM) are a vascular malformation of venous-mixed type. A small subset of CM may be associated with high flow arterial malformations, the vein of Galen aneurysmal malformations (VGAM), and have been reported to be due to RASA1 mutations.

**Aim** To report the clinical presentation and outcomes of the first two UK patients with VGAM and RASA1 mutations.

**Results** Patient 1 presented at 13 months with high grade intracranial arterial shunting, a left AV fistula, and a right AV malformation. He was treated surgically and is now well. Patient 2 presented at 6 months with a left AV fistula, right AV malformation and a right infarct. He was treated surgically and has improved.

**Conclusion** VGAM and RASA1 mutations are rare but serious vascular malformations.

**REFERENCE**

1. Laurence M Boon, Nicole Revenu, Mikk a Vikkula, Université catholique de Louvain, Brussels, Belgium.

**G23** EDUCATION AND TRAINING USING AN INNOVATIVELY ADAPTED MANIKIN: SIMPLE, AFFORDABLE, FEASIBLE AND EFFECTIVE (SAFE)

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**Introduction** Hi-fidelity manikins are often used in simulation courses. However, they are very expensive and some of the skills like drainage of pneumothorax or insertion of chest-drains/rectal probes cannot be demonstrated on these manikins as they are fully loaded with various electronic equipment inside them and puncturing will damage these expensive manikins. Hence our team developed a multi-purpose, low cost, low-fidelity manikin’s set of training modules for trainees to undertake practical skills like emergency needle thoracocentesis, pigtail chest-drain insertion, umbilical lines insertion/sampling. This method was only used for 42% of handovers. Majority (70%) of the handovers were conducted with the aid of printed sheets, which included: patient demographics (83%), presenting complaints (85%), investigations, results and treatment plans (83%). Only 11% of handovers were done electronically. Handovers had allocated start times (96%) with designated places (89%) close to area of work. However only 65% of the handovers started on time, 20% were free from distractions by allied professionals and just 5% were ‘bleep’ free. 68% had some educational activity within the time allocated in the handover. WPBAs were initiated or completed in only 11% of handovers. Overall 91% of trainees felt that the quality of handover was either average or good.

**Conclusions** The findings from our survey suggest that the quality of handovers is variable. Handovers should have a structured approach and free from distractions to ensure safety and continuity of care. Incorporating formal teaching and WPBA’s could help develop the role of handovers.

**Aims** Aim was not only to create simulation of real clinical situations but also to teach practical skills and build the concept of team working. ALS Manikin was modified as below:

1. An innovatively-designed container with red fluid was placed in abdominal cavity and connected to synthetic umbilical cord. Umbilical arterial line was connected through an innovatively-designed simulator transducer box producing arterial wave form with feasibility to vary BP using solenoid valve.

2. Manikin’s chest was drilled between ribs and lungs were made from Nitrile gloves. These lungs on connecting to flow metre were able to show positive trans-illumination test and provided air filled lungs for needle thoracocentesis and chest-drain insertion.

3. Manikin’s bottom was drilled for rectal probe insertion. Thermistor from rectal probe was removed and connexion made to an innovatively-designed simulator transducer box producing arterial wave form with feasibility to vary BP using solenoid valve.

**Following above adaptations, regular simulation sessions were initiated for:**

1. Trainees to undertake practical skills like emergency needle thoracocentesis, pigtail chest drainage insertion, umbilical lines insertion/sampling.

2. Train nursing staff with rectal probe insertion, familiarise with connexions of chest-drain and umbilical lines.

3. Both medical and nursing staff to work in team to develop effective communication.

**Background** Hereditary Hemorrhagic telangiectasia (HHT) tends to be the first condition to be considered in the differential diagnosis of patients presenting with high flow vascular malformations in combination with cutaneous vascular lesions. However, particularly in the paediatric population, capillary malformation-arterial venous malformation syndrome (CM-AVM) due to RASA-1 mutation is more likely.

**Aims** To present the clinical features of three patients with CM-AVM syndrome, promote knowledge of this condition and aid prompt diagnosis.

**Methods** Clinical examination, detailed family history, imaging (ultrasound, MRI, angiography) and genetic testing.

**Results** Patient 1 was born with a large vascular mass affecting the right side of the face and multiple cutaneous capillary malformations. Patient 2 had a spinal AV fistula and two vascular stains. Patient 3 presented with an intracranial haemorrhage secondary to a parietal AVM and was noted to have several cutaneous vascular lesions. Patients 2 and 3 were referred to the dermatology team as suspected HHT. The cutaneous vascular lesions present in all three patients were consistent with capillary malformations (in keeping with a diagnosis of CM-AVM) and were not typical of telangiectases.

**Conclusion** In patients with high flow CNS vascular lesions, it is crucial to establish the precise nature of cutaneous vascular lesions in order to request appropriate genetic testing and screening of relatives.

**REFERENCE**

1. Laurence M Boon, Nicole Revenu, Mikk a Vikkula, Université catholique de Louvain, Brussels, Belgium.
Aims Vein of Galen arterial malformation (VGAM) is a rare high flow cerebral arteriovenous malformation which most commonly presents with cardiac failure in infancy. VGAM is considered to be a sporadic disorder, with a population incidence of 1 in 100,000. No genetic basis or increased risk of recurrence within affected families has been identified previously. Recently, RASA1 gene mutations have been identified as causative in the autosomal dominant capillary malformation arteriovenous (AV) malformation (CM-AVM) syndrome, a condition presenting with multiple skin AV malformations. A large European study of affected kindreds identified associated non-cutaneous AV malformations and, amongst 140 individuals, identified two cases of VGAM, raising the possibility of a genetic basis for this condition. The aim of the present study was to assess the frequency and type of RASA1 mutations in a population presenting with VGAM malformation.

Methods A National Centre for VGAM treatment obtained consent for RASA 1 mutation analysis for all cases presented to the service from January 2011. Genomic DNA was obtained from blood samples and the 25 exons of the RASA1 gene were sequenced for each patient.

Results RASA1 analysis has been undertaken for 11 cases and four were positive for mutations: c.2912T > C (missense), c.2125C > T (truncated) and C.2119C > T (missense) (two cases). The two cases with the C.2119C > T mutation were siblings. One case, with the c.2125C > T mutation, developed the typical CM-AVM rash.

Conclusions RASA1 mutations are strongly associated with VGAM and are biologically plausible causative mutations. The autosomal dominant inheritance of this mutation, has a significant implication for counselling affected families.

REFERENCE