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VON WILLEBRAND DISEASE RECLASSIFICATION IN A NATIONAL PAEDIATRIC COMPREHENSIVE CARE CENTRE

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In 2008, the National Institutes of Health published guidelines (www.nhlbi.nih.gov/health-pro/guidelines/current/von-willebrand-guidelines/full-report/4-management-of-vwd.htm) in which the suggested level for the designation of type 1 VWD was a VWF:Ag or VWF:RCo level of <30 IU/dL and 'low VWF levels' refer to individuals with VWF:Ag levels between 30 and 50 IU/dL. The aim was to reclassify the VWD patient cohort at the National Paediatric Comprehensive Care Centre Our Lady's Children's Hospital Crumlin, Dublin.

Methods 315 Case records of children <18 years with VWD or possible VWD over a 10 year period were retrospectively extracted from the Irish National Bleeding Disorder database. These records were 'interpreted' according to NHLBI/NIH diagnostic criteria. The algorithm applied was; VWF levels <30IU/dl on 2 separate occasions–VWD; VWF levels 30–50IU/dl on 2 separate occasions–Low VWF; VWF not less than 50IU/dl on 2 occasions and multiple testing–not VWD. Blood group was also recorded. Where incomplete laboratory data, patients were recalled to a review clinic for further testing.

Results 315 children on the Irish National Bleeding disorder database had been historically diagnosed with VWD or possible VWD.

Following the review there was a 81% reduction in the number of patients diagnosed with Type 1 VWD (187 patients in the original cohort were classified as Type 1 which reduced to 36 post -reclassification).

Predictably, no significant change in numbers diagnosed with Type 2 (26) and Type 3 (2 patients) VWD,

185 (59% of total population) are now classified as Low VWF with a preponderance of Group O patients.

37 (15% of total population) were deemed to have no form of VWD and reclassified as normal.

19 (6% of total population) have not returned for full reclassification and still remain as unspecified or possible.

Discussion/Conclusion Our data suggest over-diagnosis of VWD in this population using previous guidelines. Diagnosis, especially for individuals with mildly decreased VWF (30–50IU/dl) requires correlation of clinical assessment and laboratory results. Reclassification has resulted in reallocation of resources to priority patients.

This recommendation does not preclude the diagnosis of VWD in individuals with VWF:RCo of 30–50 IU/dl if there is supporting clinical and/or family evidence for VWD or the use of agents to increase VWF levels where VWF:RCo is 30–50IU/dl and may be at risk for bleeding.

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MORBIDITY AND MORTALITY OF MEDICAL AND SURGICAL NECROTISING ENTEROCOLITIS

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Introduction Necrotising enterocolitis (NEC) is the most serious and frequent gastrointestinal emergency in the neonatal intensive care unit, (NICU) and is a major cause of death in premature infants. It is also associated with considerable morbidity, including increased respiratory support and level of care, need for intravenous antibiotics and total parenteral nutrition (TPN), and has long-term effects on growth and neurodevelopment.

Aim This study aims to evaluate the difference in mortality and short-term morbidity between infants with medically and surgically treated NEC in a tertiary-level surgical neonatal unit in Cambridge, England.

Methods This retrospective analysis of prospectively collected data evaluated infants with a diagnosis of NEC between 1st January 2009 and 31st December 2011. Diagnosis was made using modified Bells criteria and infants were defined into two groups by the treatment received. Medical NEC was treated with standard therapy of 7 days of broad spectrum intravenous antibiotics and withholding of enteral feeding, while surgical NEC was defined as the requirement for operative intervention; either laparotomy or placement of a peritoneal drain.

Results During the 3-year study period, 152 infants were diagnosed with NEC and met inclusion criteria. Of these, 82 required medical management only and 70 required surgical intervention in addition to the standard medical treatment. There was a significant difference in mortality between the two groups with survival in the medically managed group of 96% vs. 61% in the operative group ($p \leq 0.0001$). The surgical group had a statistically significant higher rate of NEC recurrence (14% versus 28.5%, $p = 0.03$). The surgical group had more days of ventilation (7 vs 13.5, $p = 0.001$), more days on TPN (26 vs 46 $p < 0.0001$) and were more likely to receive surgical central line insertion under general anaesthetic (11% vs 26%, $p = 0.02$). There were no significant differences in gestation or birth weight, type of feeding or early somatic growth.

Conclusion This is the largest single centre study comparing these two treatment groups and provides accurate contemporaneous data with which to counsel families. Operative NEC is associated with greater mortality and a higher rate of recurrence when compared with medical NEC. There is also significant morbidity associated with surgical NEC, including longer ventilation and long-term use of total parenteral nutrition, which has associated complications and sequelae.

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MULTIMODAL MONITORING AS PREDICTOR OF BRAIN INJURY IN THE PRETERM INFANT

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Background Cerebral auto-regulation(CAR) in the preterm infant is a complex, multi-factorial process which is still poorly characterised in very preterm infants. It plays a substantial role in the aetiology of intraventricular haemorrhage (IVH). Studies typically examine the relationship between cerebral oxygenation measured using near-infrared spectroscopy (NIRS) and mean arterial blood pressure(MABP) as a method to study CAR impairment.

Objective To explore a more comprehensive, multi-modal method of analysis for CAR that incorporates other potentially important factors such as continuous electroencephalogram (cEEG) and cardiac output(CO) measurements.