

mother. The patient had no additional features in his medical and family history. Physical examination revealed a weight of 4020 g (50–90p), a height of 52 cm (50–90p), a head circumference of 37 cm (90p), blueberry muffin rash (figure 1) on the trunk and proximal extremities, hepatosplenomegaly. Laboratory studies revealed a white blood cell count of  $1048 \times 10^9/L$ , hemoglobin of 8.5 g/dL, and platelet count of  $49.6 \times 10^9/L$ . Diffuse blastic cells were observed in the microscopic examination of the peripheral blood smear.

**Results** Erythrocyte and platelet transfusions were performed. The patient underwent leukapheresis once, and the white blood cell count was reduced to  $325 \times 10^9/L$ . The B-acute lymphoblastic leukemia was detected in the patient according to diagnostic tests. The 11q23 translocation was detected positive in the genetic tests. The patient was started on a chemotherapy protocol. The patient, who did not respond adequately to chemotherapy and developed septic shock and respiratory distress during follow-up, died at the age of 55 days.

**Conclusions** Congenital leukemia is a disease that is rare in the first month of life and has a poor prognosis. Skin lesions can often be the initial symptom. Skin findings in infancy should be evaluated from this perspective.

**PP-089** **ATYPICAL HARLEQUIN COLOR CHANGE BETWEEN UPPER AND LOWER HALVES BODY CASE REPORT**

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10.1136/bmjpo-2024-EPAC.233

**Aim**

**Material and Method** HCC of the newborn, known also as Intractable vascular autonomic dysregulation (Harlequin phenomenon), is a transient unilateral erythema that is seen in up to 10% of healthy newborns and was first described by Neligan and Strang in 1952. HCC is macular and blanchable displaying a sharp midline cutoff. It is often gravity dependent. HCC typically presents between 2nd and 5th day of life, but some infants may present with episodes up to the third week of life. We herein report a case of full term male who had Harlequin Color Change between upper and lower halves of his body.

**Results** 39+1w singleton pregnancy boy born by spontaneous vaginal delivery without any antenatal, intranatal issues had a bluish lower half of the body and pale upper half which lasted 4 hours and resolved spontaneously without any other symptoms. In 3rd week of life he presented to the hospital with virus infection, bluish discoloration was noted around the umbilicus and abdomen distension. ECG, ECHO, CSF analysis and abdominal ultrasound were all normal. Patient was discharged home and presented later for troponin level follow-up which was normal. The differential diagnosis includes port-wine stain, nascent hemangioma of infancy, cutis marmorata, cutis marmorata telangiectatica congenita, CHILD syndrome, Acrocyanosis, Drug reaction.

**Conclusions** The exact etiology of HCC is unknown, but the evidence points to sympathetic autonomic dysfunction in the control of the capillary tone, most likely secondary to hypothalamic immaturity in the newborns. Most cases are idiopathic in healthy newborns. HCC has also been noted in the setting of prematurity, low birth weight, hypoxia, infusion of prostaglandin E1 (PGE1), neonatal meningitis. Learning points: Harlequin phenomenon occurs in healthy newborn.

Recognition and education of this skin disorder are important as HCC is a surprising and alarming occurrence for parents, so reassurance about the benign nature of the condition, and to avoid additional diagnostic procedures and unnecessary treatments. Beyond the group age the search for an underlying cause should be vigorously pursued.

**PP-090** **BRAIN-LUNG-THYROID SYNDROME: NEONATAL PHENOTYPE OF DE-NOVO NKX2.1-RELATED DISORDER**

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10.1136/bmjpo-2024-EPAC.234

**Aim** We present a rare, genetically heterogenous, de-novo NKX2.1-related disorder in a neonate

**Material and Method** Brain-lung-thyroid syndrome, is a rare genetic syndrome.1 It is caused by autosomal dominant mutations in the NKX2-1 gene (previously TTF-1), on chromosome 14q13, which encodes for a particular transcription factor, critical for the development of the brain, lung and thyroid.2 It is a genetically heterogeneous condition, with over 30 different causative mutations identified.3 The syndrome is also referred to recently as NKX2.1-related disorders, due to the varied manifestations noted in heterozygous mutations, including benign hereditary chorea [BHC] which is a hallmark feature, while some can develop choreoathetosis, tremors, motor & vocal tics as well.4 Classic triad of the syndrome includes Congenital hypothyroidism, Benign hereditary chorea and Respiratory distress syndrome in infancy. Approximately 50% of the patients have a complete triad involved, while about 30% are affected by neurological and hypothyroidism phenotype and about 13% may only present with neurological phenotype. The overall prognosis varies depending on the presenting phenotype and severity of symptoms with respiratory involvement having the most impact on survival in some patients.5 We present a term neonate admitted in the neonatal intensive care unit for respiratory distress with incidental finding of macroglossia leading to diagnosis of congenital hypothyroidism with hypertonia at birth, with childhood Interstitial lung disease [figure 1]; eventually diagnosed as heterozygous for the pathogenic deletion of the entire coding region of NKX2-1 gene including the promoter region, de-novo mutation.



**Abstract PP-090 Figure 1** CT Thorax: Ground glass appearance suggestive of childhood interstitial lung disease

**Results** We present a term neonate admitted to the neonatal intensive care unit for respiratory distress with incidental finding of macroglossia leading to diagnosis of congenital hypothyroidism with hypertonia at birth, eventually diagnosed as heterozygous for the pathogenic deletion of the entire coding region of NKX2-1 gene including the promoter region, de-novo mutation.

**Conclusions** High index of suspicion is required in neonates with congenital hypothyroidism and respiratory distress syndrome to determine an underlying genetic etiology. Life expectancy in individuals with NKX2-1-related disorders is expected to be normal with supportive management.

**PP-091 PERCEPTION TOWARD THE NEED FOR MORPHINE IN VENTILATED PRETERM INFANTS: A COMPARATIVE STUDY BETWEEN A TERTIARY NEONATAL UNIT AND THE OPERATIONAL DELIVERY NETWORK (NWNODN)**

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10.1136/bmjpo-2024-EPAC.235

**Aim** The routine administration of morphine to sedate premature ventilated infants is not recommended by professional organisations such as NICE. However, the routine use of morphine persists due to perceived pain and a lack of awareness of non-pharmacological pain management. Additionally, morphine is associated with risks such as IVH, prolonged ventilation, and low blood pressure. As part of a quality improvement initiative, we surveyed staff in our unit and the Northwest Neonatal Operation Delivery Network (NWNODN). This study aims to compare perceptions regarding the use of morphine in ventilated premature infants between our unit and the rest of the network.

**Material and Method** An email survey was distributed to all staff in our neonatal unit from July 15th to August 5th, 2022. The same survey, with minor adjustments to accommodate the network’s diversity, was sent to the Northwest

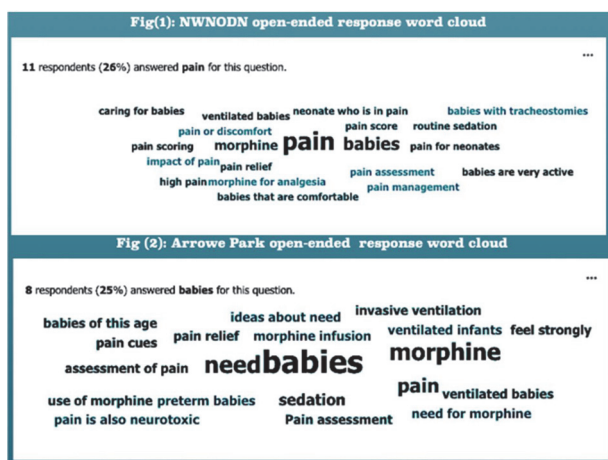
**Abstract PP-091 Table 1** Comparing the perception, knowledge, and practice between the unit and the network

|   | Arrowe Park Hospital | NWNODN         |
|---|----------------------|----------------|
| <b>Respondents</b>  | 32                   | 42             |
| <b>Is it routine in your unit to intubate infants born &lt; 32 weeks at birth in the Delivery room or Operation Room?</b> | Yes 8 (25%)          | Yes 9 (21.4%)  |
| <b>Do you routinely start them on morphine infusion when they are mechanically ventilated?</b>                            | Yes 10 (31%)         | Yes 12 (28.5%) |
| <b>Do you use any method or scoring system to assess pain in premature infants?</b>                                       | Yes 31 (97%)         | Yes 39 (93%)   |
| <b>Have you heard about the N-PASS pain assessment tool?</b>  | Yes 25 (78%)         | Yes 21 (50%)   |
| <b>Do you use the N-PASS in your unit?</b>  | Yes 21 (66%)         | Yes 8 (19%)    |
| <b>During clinical rounds, is pain scores part of the clinical presentation?</b>  | Yes 6 (19%)          | Yes 17 (40%)   |
| <b>Does your unit have guidelines on how to assess and manage analgesia/sedation during invasive ventilation?</b>         | Yes 19 (59%)         | Yes 29 (69%)   |
| <b>Do you think that preterm infants need sedation/analgesia while on invasive mechanical ventilation?</b>                | Yes 18 (56%)         | Yes 27 (64%)   |
| <b>Have you heard about NIDCAP?</b>   | Yes 12 (38%)         | Yes 24 (57%)   |
| <b>Is NIDCAP part of the practice in your unit?</b>   | Yes 2 (6%)           | Yes 3 (7%)     |
| <b>Aware of the National and International recommendations?</b>   | Yes 19 (59%)         | Yes 19 (45%)   |

Neonatal Operation Delivery Network (NWNODN). The survey was disseminated via the weekly Network bulletin, and responses were collected between October 6th and November 14th, 2022. Descriptive and thematic analyses were employed to analyze the results.

**Results** There were 32 and 42 respondents from our unit and the NWNODN, respectively. The majority of respondents were Registered Nurses (RN) (APH 50%, NWNODN 45%) and were from level 3 tertiary units (64%). Perceptions regarding morphine and pain management in ventilated premature infants were similar between APH and NWNODN (table 1). More than half of the respondents believed that ventilated premature infants require morphine during ventilation (APH 56%, NWNODN 64%). The most frequently used words by APH staff were ‘Babies,’ ‘need,’ and ‘morphine,’ while the NWNODN staff most commonly used ‘Pain,’ ‘babies,’ and ‘morphine’ see figure 1.

**Conclusions** There were no differences in the perception of pain and the use of morphine in ventilated premature infants between APH and the network. However, this survey identified a knowledge gap and attitude toward pain management in ventilated premature infants.



**Abstract PP-091 Figure 1** NWNODN open-ended response word cloud.