GP272 TRENDS IN THE MANAGEMENT AND OUTCOMES OF BABIES VENTILATED FOR MECONIUM ASPIRATION SYNDROME IN A TERTIARY NEONATAL INTENSIVE CARE UNIT

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Background Meconium aspiration syndrome remains a condition still encountered in the neonatal intensive care units resulting in significant morbidity and mortality among term neonates. With advances in neonatal intensive care management, the outcomes of babies with meconium aspiration syndrome have improved significantly. These include the use of high frequency ventilation with inhaled nitric oxide. This has significantly reduced the need to send these babies for extracorporeal membrane oxygenation (ECMO) treatment.

Aims To review the management and outcomes of babies ventilated for meconium aspiration syndrome in a tertiary neonatal intensive care unit (NICU).

Methods We retrospectively searched the Badger electronic patient record for all babies with a diagnosis of meconium aspiration syndrome in the last 9 years between 01/01/2010 and 31/12/18. We then selected all ventilated babies with meconium aspiration syndrome for this review. A structured excel spread sheet was used to collate and analyse the data.

Results In the last 9 years there were 77 ventilated babies who had a diagnosis of meconium aspiration syndrome assigned to their badger electronic patient record. 59 were inborn and 18 were outborn babies. The median gestation age was 40 weeks with a range between 35 and 42 weeks. The median birth weight was 3470 grams with a range between 1836 grams and 4850 grams. 47 babies were managed with conventional ventilation and 30 babies were on high frequency ventilation. 38 babies received surfactant as part of their treatment for meconium aspiration syndrome. 37 babies also received inhaled nitric oxide. 43 babies had inotropic support. 3 babies had ECMO. One was an inborn baby referred for ECMO in 2014. The other two were out-born babies transferred back to the NICU following ECMO for on-going management. All 3 babies who went for ECMO were discharged home. 2 babies died in the last 9 years (2014 and 2015) but these were attributed to their neurological conditions.

Conclusion The outcomes of babies ventilated for meconium aspiration syndrome are good despite the serious nature of the disease. Swift escalation of the management of persistent pulmonary hypertension of the newborn (PPHN) is the key to reducing morbidity and mortality. With advances in the management of PPHN, we had only referred one inborn baby for ECMO in the last 9 years for meconium aspiration syndrome. The results of this review provide guidance when counselling these traumatised parents of babies ventilated for meconium aspiration syndrome.

GP273 CORKAMBI: ONE YEAR OUTCOMES OF ORKAMBI ON CYSTIC FIBROSIS PAEDIATRIC PATIENTS IN CORK UNIVERSITY HOSPITAL

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Introduction Orkambi (Lumacaftor/Ivacaftor) was first introduced for use in the Cystic Fibrosis population in Ireland in 2017. It is only licensed for those patients who are homozygous for the F508del gene mutation. Orkambi was originally approved for children aged 12 years and older in 2017 and then this was extended for those aged 6–11 years old in 2018. Recently, the EMA (European Medicines Agency) has approved use for children 2–5 years. CUH (Cork University Hospital) looks after 98 paediatric patients with CF of whom 37 have been commenced on Orkambi since July 2017.

Objectives The primary objective was to assess the outcomes of Orkambi at 6 and 12 months on FEV1, BMI and BMI Centile and to compare the days on Intravenous antibiotics and weeks of oral antibiotics for pulmonary exacerbations in the year before and after commencing Orkambi.

Results Thirty seven patients were included in the study population with a split of 21 females and 16 males. The average profile of our patient cohort before commencing Orkambi was 11.0 years, BMI 16.8 kg/m², BMI Centile 43.2 and Average FEV1 96.7%. To date, 11 patients have completed over a year of treatment and the remainder will have completed 12 months by June 2019. Six month followup has shown that 30/37 (81%) of patients have had an increase in their BMI with an average BMI at 6 months of 17.5 and BMI difference of +0.7 kg/m². BMI centile was increased in 24/37 (65%) with centile difference of +5.7 to an average of 48.9. Lung function did not show any improvement from Pre-Orkambi values with average FEV1 of 95.9% at 6 months. This was also shown in followup of patients who completed 12 months of treatment with average FEV1 difference of -2%. For those who had completed 12 months on Orkambi, the no of weeks on oral antibiotics and IV antibiotics both showed a reduction in the year following commencement. There was a 26% decrease in oral antibiotics prescribed with average reduction of 1.5 weeks. Three patients had required IV antibiotics in the year prior. Two of these had no IVs over the following year and one patient with severe CF disease had a 55% reduction in days on IVs from 121 to 66 days.

Conclusion Orkambi has improved BMI/BMI centiles but has not shown any improvement in FEV1. Preliminary data has shown a reduction in pulmonary exacerbations although further analysis of the remaining patients will allow more accurate conclusions.

GP274 LIVER TESTS IN F508DEL HOMOZYGOUS CYSTIC FIBROSIS PATIENTS ON ORKAMBI

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Aims Cystic Fibrosis (CF) is an inherited condition causing complex, multi-system disease. Ireland has the highest incidence of CF in the world, where one in 19 people carry a defective gene. The most common detected Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) mutation in Ireland is the F508del. In 2016, 55% of patients were homozygous for F508del. Orkambi is a combination of Lumacaftor and Ivacaftor, and is licensed in the treatment of F508del homozygous CF patients. In patients taking Orkambi, serious adverse reactions related to elevated liver transaminase levels have been reported. It is recommended that ALT, AST and bilirubin are monitored every three months for the first year

of treatment and annually thereafter. Our aim was to determine if patients taking Orkambi experienced a rise in their liver enzymes during treatment.

Methods This study was conducted at the University Hospital Limerick, Ireland Paediatric CF unit. All 39 homozygous F508del patients on Orkambi were included. Lab results were collected for the following variables for each patient: AST, ALT, ALP, GGT and bilirubin at set time intervals Pre-Orkambi, and three, six, nine and 12 months on treatment. The Pre-Orkambi levels were compared with the three month levels.

Results We determined that overall patients on Orkambi did not have a rise in liver enzymes. Unexpectedly, patients had a statistically significant decrease in their ALT (p-value 0.016), ALP (p-value 0.001), GGT (p-value 0.002) and Bilirubin (p-value 0.002). AST levels saw no significant change (p-value 0.76). Two patients had a rise in AST levels during Orkambi treatment greater than three times ULN, however, these levels then decreased with continued Orkambi use. None of our patients had any liver tests greater than eight times the ULN, or five times ULN during treatment, and none had ALT, ALP or Total Bilirubin levels greater than three times ULN during treatment. None of our patients had to stop taking Orkambi due to adverse events related to liver tests. Our findings are in contrast to those suggested by the original Orkambi trials data

Conclusion Our study demonstrated that patients taking Orkambi do not experience a rise in liver tests, in fact, unexpectedly, the values decrease.

GP275 PEDIATRIC SPRAY-SOL: A NEW NEBULIZATION DEVICE SPECIFIC FOR UPPER RESPIRATORY TRACT INFECTIONS

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Background Upper respiratory tract infections (URTI) are very common in the pediatric population due to their 'physiologically' immature immune system; yet, around 90% are viral – only 10% of URTI are bacterial. Therefore, treatment should essentially be symptomatic to relieve the child's discomfort and symptoms. Topical, rather than system, treatment is preferable reducing systemic effects and increasing the topical efficacy. There are many different nasal devices used to treat URTI, but often they are not specifically targeted for this purpose.

Objective Our group developed a new nebulization device, called 'Pediatric spray-sol', target specificity, very speed, pocked-size and portable, mono-patient without sterilization problems with the ability to vary concentration and quantity of medication in delivery, to combine substances in a single treatment.

Materials and Methods The device can be used in combination with a common syringe (without needle), without need for electricity. The user operates the plunger of the syringe to push the liquid drug into the delivery device. Once inside, the substance is nebulized and, from there, delivered into the nose in aerosol form. The device includes a fluidodynamic path with an expansion chamber and an acceleration chamber

arranged in sequence, which allows for the optimum delivery of fluid.

Results The features of Spray- Sol include: the ability to nebulize high viscosity substances similarly an aerosol, nebulize particles of average diameter between 10 and 20 microns, the very low administration times (10 seconds to 5 cc of substance), the cost and the possibility of use for all the duration of the treatment, it is portable and doesn't need electricity. This nasal nebulizer provides a more extensive and intensive delivery of solution in the nasal cavities, especially the deeper and higher portions of nasal cavity. In particular, pediatric Spray-sol, seem to guarantee a homogeneous distribution of drug at the middle turbinate and ostiomeatal complex crucial region for the drainage and ventilation of the paranasal sinuses. Spray-Sol reaches more easily the nasopharynx subsite and it could be considered a useful device for adenoiditis and Eustachian tube dysfunction. Moreover, it can quickly and efficiently nebulize not only saline solutions but others substances like hyaluronic acid, ectoine and corticosteroids.

Conclusions Pediatric Spray-sol is a new nebulization device for URTI treatment: combining the comfort of a spray with the efficacy of an aerosol. Pediatric Spray-sol can be used to deliver any type of solution in the nose such as saline solutions, hyaluronic acid, ectoine and corticosteroids.

GP276 AUDIT OF NEWBORN SCREENING CORK UNIVERSITY HOSPITAL 2011–2019 – IMPACT OF GESTATIONAL AGE ON SWEAT TESTING

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Background A previous national audit looking at sweat testing in all age groups in 2011 found Cork University Hospital (CUH) had a high number of tests with quantity not sufficient (QNS) result. Since then there has been new equipment and staff training. International laboratory audit criteria examine failure rates for under 6 months which has recently been <1% in CUH (acceptable<5%). However higher failure rates (0–40%) for sweat testing have been noted internationally in newborn screening programmes, a more difficult cohort to successfully sweat test.

Aims Sweat tests performed in CUH from 2011–2018 as part of newborn screening in relation to number of patients with positive (>60), negative (<30), equivocal and QNS results were reviewed. The gestational age at birth and at time of testing and birth weight were examined as potential variables.

Methods

Ethical approval obtained Newborn screening records for all patients from July 2011 to December 2018 reviewed. Sweat testing data obtained from laboratory. Data analysed using SPSS and Chi-Square test.

Results 102 infants referred to Cork for sweat testing as part of newborn screening. Patients diagnosed by meconium ileus (n=7), had sweat testing elsewhere (n=1) or had antenatal diagnosis (n=2) were not included (N= 10). On initial testing 30 patients had a QNS result (32.6%). 7 had same day repeat testing which was sufficient, the remaining having sweat testing at a later date. Ultimately there were 23 with positive tests, 67 with negative tests, 2 equivocal tests. Gestational age and birth weight of the 30 patients with initial QNS result