

Changing Population and Treatment of Children and Young People with Cystic Fibrosis in Post New-born Screening Era a Large Shared Care District General Perspective

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Aim : Comparison of children and young people with Cystic Fibrosis [CF] attending a large district shared care centre pre and post new-born screening [NBS].

Methods: A local regional audit of CF patients in 2005 [n=37] NBS and 2017 [n=38] post NBS were compared to look for trends which maybe useful for future service development.

Results: Comparison of the 2005 versus [v] 2017 cohorts showed: Mean ages 9 v10 years

Presentation of CF: NBS 0% v 39%, Respiratory symptoms 61% v 30 %, Failure to thrive 47% v 39 %, Meconium ileus 20% v 18%.

Screen positive Indeterminate diagnosis 0% v 5%.

Total cultures of Pseudomonas Aerginosa Psd Ag] 27% v 21%, Staphylococcus aureus 17% v 7%, Non-tuberculous mycobacterium[NTM] 0% v 1% ,Aspergillus [Asg]4% v 6%, Gram negatives 4% v 11%, Haemophilus Influenzae 10% v 3 %, Serratia Marscens 0% v 2%.

Treatment : regular intravenous antibiotics 38% v 15%, nebulised antibiotic 44% v 17%, central lines / porta-caths 36 % v 18 %;DNase 30% v 39%, CFTR potentiator Rx 0% v 8%, Ursodeoxycholic acid 0%v 13 %. **Complications :** Liver disease 9% v 13 % , Diabetes 8% v 13 %.

NBS accounted for over a third of 2017 cohort with increasing number of SPIDs whose prognosis differs from the classic CF patients. NBS reduces the number of symptoms at diagnosis and total cultures in 2017 population has shown a decrease in Psd Ag and SA but an increase in gram negatives, Asg and NTM. Treatment differences correlate with the decrease in Psd Ag isolates with less use of regular IVs, portacaths and nebulised antibiotic Rx. Liver disease and CF Diabetes is more common perhaps due to better surveillance.

Conclusion: This comparative audit gives useful data to inform local service development eg SPID clinic and infection control.