Developing a specialist nurse run adult sweat test clinic

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Background: CF diagnoses are increasingly made in adulthood. Sweat tests (SWT) are routine in paediatrics but less common in adult medicine. A telephone survey of UK adult CF centres showed that one out of 16 held a diagnostic SWT clinic. All others performed SWT on an ad hoc basis by biochemists (n = 9), paediatric teams (n = 4) and clinical nurse specialists (CNS, n = 3).

Objectives: To audit a newly introduced CF CNS run formal SWT clinic at the Royal Brompton Hospital (RBH) Adult CF Unit. Three SWT clinics are held per month. The Wescor Macropust test system is used and national guidelines followed.

Results: The service has received 83 referrals over 25 months (males 49%; median age (range): 36 yrs (16–76 yrs); 18% (n = 15) did not attend appointments). Referrals were received from: general practitioners n = 16 (19%) and other hospitals n = 31 (37%); other referrals were from non-CF RBH colleagues. Patients had 2 simultaneous tests (total 136 tests). 3% (n = 4) tests were unsuccessful (x1 SWT on two patients due to insufficient volume and x2 SWT on one patient who interfered with collection). 65% (n = 44) had a sweat chloride >30 mmol/L, 6% (n = 4) had a chloride ≤30 mmol/L. After testing patients are further evaluated in the Difficult CF Diagnosis clinic. Additionally patients (n = 19) with the G551D mutation on sweat testing were referred to the CF CNS for additional management.

Discussion: This audit shows that a CNS run adult SWT clinic is a successful service. It serves both the patient care and education needs of patients and staff. It demonstrates the ethos of a “CF in adult medicine” approach.

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Lung infections in patients with cystic fibrosis – leading microbial pathogens

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Objectives: The aim of the study was to determine the spectrum and susceptibility to antimicrobial agents of the microorganisms, isolated from respiratory tract (RT) in patients with cystic fibrosis.

Methods: The study includes 25 patients aged from 3 months to 32 years, treated at UMHAT Pleven between 2009 and 2014. A total of 75 clinical specimens obtained from the RT were analyzed. Identification and susceptibility testing were performed by conventional methods, VITEK 2 and mini API Systems.

Results: Bacterial findings were registered in 46 samples from 21 of the patients. A total of 56 microbial isolates were cultured, comprising 42 clinical strains. Gram(−) bacteria were predominant – 66.7%, followed by Gram(+) bacteria – 28.6%, and yeasts – 4.7%. Among Gram(−) bacteria non-fermenting glucose rods are the most common organisms detected – 8.6%: P. aeruginosa 17 strains, P. alcaligenes 2, A. faecalis 1, A. xylosidans 1, O. anthropi 1. P. aeruginosa strains were isolated from 13 patients of different ages, but the majority of patients were over 5 years of age. P. aeruginosa showed good susceptibility to antimicrobial agents. All isolated strains were susceptible to tobramycin, cefazidime and meropenem, but had some phenotype characteristics that highly distinguished them. 13/17 strains were of mucoid phenotype. This phenotype caused 3 out of 4 deaths in the study group.

Conclusion: Microorganisms isolated from RT varies and changes when monitored for long time. Gram(+) bacteria, especially P. aeruginosa are dominant. P. aeruginosa showed unusual phenotype characteristics.

Reducing the rate of inadequate sweat testing for the NBS in the state of Michigan

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Objectives: In Michigan (MI), cystic fibrosis (CF) Centers-specific QNS rates ranged from 12–25% in 2009 instead of the 10% rate recommended by the CF Foundation (CFF). That led to delayed diagnosis, psychological and financial burdens on families. In this study, 4 of 5 CF Centers in MI actively participated but all five worked together on strategies to reduce QNS rates.

Methods: MI CF Centers in addition to the Michigan Department of Community Health (MDCH) collaborated on this project. Project steps:
1. Review the current sweat testing methods and protocols employed by participating centers.
2. Invite an expert in the sweat testing field to each center to review the sweat testing process. A detailed report was sent to each center with recommendations and suggestions to improve the technique, quality of sweat collection, analysis and running of the test.
3. Sharing of the information between the CF Centers through visits by the sweat testing personnel to share knowledge and help strengthen the relationship between centers.
This QI project is unique since it included several strategies to address the problem in addition to including all centers in MI. Study duration was 2 years.

Results: 778 infants were identified as positive for CF screening through the NBS Program. The mean age at time of sweat test was 23.2 days with SD of 13.0 days and median of 20 days. The overall QNS percent decreased from 14.4% to 9.5% (p-value < 0.04) during the duration of the study.

Conclusion: This project provided all participating MI Centers the opportunity to collaborate and to improve a common problem. This teamwork led to a significant decrease of sweat test QNS rates.

Feasibility of 1H-NMR spectroscopy to detect occurrence of Pseudomonas aeruginosa in saliva of cystic fibrosis patients

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Cystic Fibrosis (CF) is an important genetic disorder really common in Caucasian population. Bacterial respiratory infections are the main causes of morbidity and mortality. Pseudomonas aeruginosa is the principal pathogen of lower airways, the mainstay of antibiotic therapy. Nowadays, its diagnosis is made by culture or real-time PCR. High-resolution nuclear magnetic resonance (NMR) spectroscopy is now a fast analytical tool providing information regarding the metabolic profiles of biofluids. Saliva is an accessible biofluid for the overall health. Salivary metabolites profiling offer an interesting approach to identify biomarkers associated with several chronic disorders and cancers.

The aim of this study is to improve a protocol in order to explore the feasibility of NMR in CF, to look for intra-subject variability in salivary metabolites concentrations and to detect differences before and after Pseudomonas aeruginosa lung colonisation.

Methods: Saliva of 3 CF patients was collected. Patients haven’t eaten, drunk and tooth brushing for at least 2 hours before to take sample. Three controls saliva were obtained from healthy people. Saliva spectra were acquired by using a Bruker Avance 500 spectrometer.

Results: Exploration of salivary spectrum is under progress. We have observed a large amount of differences between samples. The protocol used for analyze samples require to be improved in order to examine modifications due to Pseudomonas aeruginosa presence.

Conclusion: Others studies have demonstrated that NMR is an efficient method to systematically evaluate bacterial infection or disease progression. In the future, NMR could provide a potentially saliva diagnostic test in CF patients.