

473 Views on medicines of adults with cystic fibrosis

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Introduction: The treatment of cystic fibrosis (CF) is complex, intrusive and often time consuming leading to difficulties with adherence. Individual patient views on medicines vary and should ideally be discussed as part of the prescribing decision. This study investigates the views on medicines of adults with CF.

Methods: The study consisted of semi-structured interviews with 25 patients attending an Adult CF Unit. Qualitative data was analysed according to the thematic Framework method as described by Richie and Spencer.

Results: Views about taking medicines ranged from 'it's a necessity' (13 patients), to 'a desire not to take but acceptance of necessity' (7), 'a desire not to take' (3) and 'a neutral attitude' (2). 80% of patients thought their medicines were beneficial or very beneficial. 72% thought that medicines were important or very important at relieving symptoms and 80% thought they were important or very important in preventing longer-term problems. Only one person described them as causing a disruption to their life, which contradicted the general way that the patients described the disruption to their daily routine. Only 4 patients raised concerns of side effects. The positive views were sometimes contradicted in the described adherence level.

The general view on the use of illegal drugs was negative with the possible exception of cannabis for multiple sclerosis or arthritis. The views on complementary therapies were varied; only 40% had some experience of using complementary therapies.

Conclusions: The views expressed on medicines varies between individuals. In some cases they had an effect on adherence and in others they did not. An individuals views on medicines should be discussed as part of the prescribing decision.

474 Doll-like face: is it an underestimated clinical presentation of cystic fibrosis?

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Cystic fibrosis (CF) is inheritable chronic disease, characterized chiefly by obstruction and infection of airways and by maldigestion. Pancreatic insufficiency is present in approximately 85% of patients with CF. Kwashiorkor is a well-recognized complication of CF characterized by hypoproteinemia, edema and anemia and can be the presenting symptom of CF. The edema of the face can be seen in kwashiorkor.

Aim: As doll-like face is very rare clinical presentation of CF patients complicated with hypoproteinemia, we evaluated demographic features and laboratory findings of 5 patients diagnosed as CF with doll-like face (DLF).

Methods: Between June 2005 and January 2008, 115 children were diagnosed as having CF enrolled in our center. 5 infants diagnosed as CF with DLF before the age of 6 months participated in the study. 48 infants diagnosed as CF without DLF before the age of 6 months participated in the study as controls (group2).

Results: The incidence of DLF less than the 6 months of age were 9.4% in our center. Physical examination revealed DLF and pitting edema of lower extremities in group 1. Their weight and length were under the third centile. Laboratory findings of group 1 include: mean hemoglobin 7.6g/dl; mean total protein 4.4 g/dl; albumin 2.3 g/dl. When compared control group in order to; 11.4 g/dl (7.6–17.9); 6.2 g/dl (4.0–8.8); 4.7 g/dl (2.1–5.8); mean hemoglobin, total protein and albumin values were significantly lower in group 1.

Conclusion: In a subgroup of patients, DLF may be the presenting manifestation of patients with CF. Especially in developing countries clinicians should be aware of in patients with malnutrition and DLF and CF should be considered in differential diagnosis.

475 A solution for severe urinary incontinence in women with cystic fibrosis: a case series

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Introduction: Urinary incontinence is more common in females with CF than in the general population. It impacts significantly on physical and psychological health. Those affected experience distress yet under-prioritise their symptoms. Standard non-operative interventions may be unsuccessful in CF and surgery has previously involved too much risk. Tension-free Vaginal Tape (TVT) is a newer, minimally invasive operative technique, popular in the general population due to improved efficacy, shorter operating time and faster recovery.

Objective: To determine whether TVT is successful and safe for women with CF and severe stress incontinence.

Methods: Four adults with CF and incontinence were formally assessed by gynaecological and urological specialists. All shared a history of years of distressing urinary leakage and no improvement despite specialist physiotherapy input. All had moderately severe CF, with multiple complications and frequent IV therapy requirements. They were admitted to our CF ward for IV antibiotics and physiotherapy to optimise their chest prior to the TVT. Ages were from 25 yrs to 47 yrs, and FEV₁ (% predicted) and BMI values ranged from 26% to 78% and 19.3 to 24.9.

Results: All patients tolerated the procedure well. Only one required a general anaesthetic. All needed only a few days stay on the ward. No patients experienced any chest deterioration after surgery. In three of them, leakage ceased completely. The fourth patient experienced considerable improvement.

Conclusions: TVT is a safe, effective and worthwhile solution for stress incontinence in women with CF. Complex CF disease should not be perceived as a barrier to this simple surgical procedure.

476 Perceived adherence of adults with cystic fibrosis and the views of the multidisciplinary team

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Introduction: The treatment of cystic fibrosis (CF) is complex, is often intrusive and may take a large portion of the day leading to difficulties in a predominantly young, mobile population. This study investigates the perceived level of adherence and factors affecting this of adults with CF and the views of the multidisciplinary team.

Methods: The study consisted of semi-structured interviews with 25 patients and ten members of the multidisciplinary team. Qualitative data was analysed according to the thematic Framework method as described by Richie and Spencer.

Results: Patients took 9 (3–17) different medicines a day with a frequency of 4 (2–6) times a day excluding the additional use of enzymes with meals, snacks and feeds. The estimated percentage adherence was 95% (10–100%) by the patients themselves. This value was contradicted by the verbal descriptions of their medicine routine by four participants in specific circumstances. The perceived adherence to individual medicines varied. The members of the multi-disciplinary team predicted different adherence rates than the patients, with a median predicted adherence level of 50% (25–60%).

Factors described that affected adherence were divided into the following themes: barriers – general, adverse effects and physical problems, motivators and facilitators.

Conclusions: The patients' descriptions of their adherence may conflict with descriptions of their daily routines involving medicines. Individual treatments should be discussed with each patient in the context of their everyday life and achievable adherence targets agreed. Perceived adherence of the multidisciplinary team differs from that of the people interviewed in this study.