Fifty years ago Cystic Fibrosis (CF) was a fatal disease of childhood. The terminology has now changed to the description of CF as a life limiting disease, or more recently a chronic lifelong disease. Recent analysis of survival data suggest that someone born with CF in 2000 is expected to live at least to the age of 50 [1]. In the context of such changes in morbidity and mortality, issues of reproductive health have gained importance in the overall scheme of managing young adults with CF.

It is recommended that young men with CF are informed that 95% of CF males are infertile because of bilateral absence of the vas deferens but that sperm aspiration and subsequent intra-cytoplasmic sperm injection can result in successful pregnancy. Thus male infertility which seemed irreversible when first described [2] is now a treatable complication.

The advice given to young women with CF has also changed dramatically in the same timeframe. Pregnancy would previously not have been possible because of amenorrhoea associated with severe end-stage pulmonary disease. Today, although the true fertility rate in CF females remains unknown, it seems that fertility is the rule rather than the exception.

It is clear that pregnancies in CF women occur and are successful but require careful planning to ensure the best outcome for mother and baby [3]. Frank Edenborough and colleagues are to be congratulated on the production of the very first set of international guidelines for the management of pregnancy in women with CF. Importantly the guidelines do not simply cover the management of the woman with CF during pregnancy but emphasise the careful discussions that should occur pre-conception regarding maternal health and longevity as well as genetic screening of the partner. The authors also include sensible guidance regarding the issues of termination of pregnancy in CF.

Discussions about pregnancy in CF often evoke vigorous debate regarding the balance between the rights of an individual to enjoy parenthood versus the possibility that a child is born destined to lose its mother before reaching adulthood. Furthermore, public health strategists may suggest that we are continuing the survival of a mutant gene resulting in further healthcare costs. Adult physicians support people with CF to achieve the best quality of life possible along with improved survival. Our success in doing so means that we are now responsible for ensuring that women with CF make informed choices about becoming a mother. The guidelines appropriately emphasise pre-conception counselling with discussion of the risks to a couple of producing a baby with CF along with discussion of the effect a pregnancy and subsequent care of a child will have on the mother.

The key to good outcomes in CF remains well co-ordinated multi-disciplinary care and the emphasis on this in the pregnancy guidelines is clear. Dieticians and physicians will find the guidance on weight gain, vitamin supplementation and when to perform and repeat the oral glucose tolerance test invaluable. The checklists in the appendix for each discipline will be sure to find their way into protocols for patient management during pregnancy, in every adult clinic.

As survival continues to improve, managing a woman with CF through pregnancy will become routine in adult CF care. These guidelines form the "mark in the sand" for future research and international collaboration to ensure that best care is established.

References


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