Patients with cystic fibrosis should not be intubated and ventilated

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Introduction

The use of pro–con debates is one of the most useful ways of learning about a difficult subject as debating a position one may or may not agree with allows one to look at the facts with a more critical eye and allows the audience to hear both sides of an issue rarely present in a guest lecture where only a single narrative argument is usually promulgated. There should be more of them.

Argument

Despite the gratifying increase in life expectancy in patients with cystic fibrosis (CF), death, as for all of us, is inevitable. A patient's preferred manner of death, if predictable, is highly personal, but medicine's, and indeed society's, increasing obsession with life at almost any price is always in danger of interfering with a dignified death. Death from CF in childhood is now rare and a CF paediatrician may not care for a child who is dying for some years. When a death does occur, all those feelings of 'I wonder if I had tried treatment X sooner or at all, then things would have been different' come to mind. Clinicians are reluctant to broach the subject of death with adults and even more so children. Sawicki[1] showed that of 234 adults with CF, 79% were happy to discuss advance directives even though only 28% had been asked by their clinicians about such issues.

It is with this in mind that the decision to institute invasive ventilation (ETV) for a severe pulmonary exacerbation in a patient known to have severe lung disease must be considered very carefully. It is important to point out what this discussion is *not* about. First, it excludes usually children presenting very ill who subsequently turn out to have CF. Second, it excludes those known to have CF but who are not severely debilitated and suddenly have an unexpected, devastating turn of events. Third, it excludes CF patients with 'non-CF' illnesses (e.g. overdoses or status epilepticus). Fourth, it excludes postsurgical ventilation for, as examples, pleurodesis or bronchial artery coiling. However, with all the above, avoiding or at least minimizing invasive ventilation and using non-invasive ventilation cannot be over-emphasized as it is potentially very useful for therapy, symptom relief, palliative care or as a bridge to transplantation but is outside the scope of this debate. Finally, like everything else, all cases must be judged at the time on their merits quite a lot of which is definable as above but some of which still distinguishes medicine from a pure science and goes on the hunch and experience of the clinicians.

My personal experience of ETV has been uniformly awful. All four severely affected children (>10 years) have died while ventilated or been switched off after discussion with the parents.

The literature is only marginally less gloomy. The original study in 1978 quoted a 94% mortality[2] but in the last few years several cohort type studies have reviewed their results. When looking at these results, one has to disentangle not only the outcomes for pulmonary exacerbations requiring ETV from the headline all-cause mortality (which is always lower), but also the difference between patient and episode numbers. In addition, if the audit covers 10–15 years there is the risk of an era effect given the rapidly changing care patterns of both CF and indeed intensive care.

Ellaffi's French single-centre (Paris) retrospective adult audit[3] covers 69 CF all-cause ICU admissions between 1997 and 2001. The overall ICU mortality was 48%, but the four cases needing ETV had a 100% mortality. Sood's review of adult ICU data from Chapel Hill, North Carolina[4] reported a

40% ETV ICU mortality rate (12/30). Eight of the 20 ETV survivors were transplanted while ET ventilated on ITU, a rare situation in the UK. However, the one year untransplanted mortality after ETV was 93%. A multicentre French cohort study reported 60 ICU admissions in 42 subjects.[5] The headline ICU mortality was only 14%, however, they identified a 16-fold increased risk of death (95% CI 4–63) with ETV. For respiratory exacerbations, overall mortality was 28%, 58% if needing ETV, and 73% if prior NIV had failed.

The only audit with a reasonable number of children is Berlinski's from Texas[6] in which they reviewed 33 subjects from age 1 month to 34 years receiving ETV for respiratory failure. There was a clear age-dependent mortality rising from 22% if <5 years through 67% if aged 5–15 years to 80% (12/15) if >15 years. Malnutrition (<80% desired weight) or a previous massive haemoptysis each significantly raised the mortality risk three-fold ($p<0.05$). Vedam's Australian review of 20 admissions[7] in those >15 years for ETV over 15 years had a 55% overall mortality rising to 100% if the adult BMI was <18 and 80% if the FEV_1 was <24%. Finally, the Dutch reported[8] that five children <2 years had zero mortality then there were no admissions for pulmonary exacerbations till age 15 years when the ETV mortality was 82% and the NIV mortality was 56%. Thus, an unweighted average of all these studies shows a 74% mortality for those aged >15 years and 67% for children.

The death of at least three-quarters of ETV patients remains very poor if slightly better than in 1978 but it remains highly dubious, at least to me, that the generally undesirable death of three patients while invasively ventilated on ICU for the marginal survival of the fourth, who will almost certainly die within 12 months if not transplanted, merits ETV for all whose gas exchange requires it. It could reasonably be argued that ETV may be required so that relatives can be gathered together 'to say goodbye' and, of course, consideration of every case on its merits is essential. Nevertheless, the need for palliative care in children remains under-recognized, certainly in the UK and indeed worldwide.[9] It behoves all CF clinicians faced with a deteriorating patient not only to maximize realistic treatments including transplantation, but to grasp the unpalatable nettle that failure and death are real possibilities, and to help the patient and their family face this as sensitively as possible.

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