Prolonged mechanical ventilation as a consequence of acute illness

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Abstract

***Objective*—To determine why acutely ill children become dependent upon mechanical ventilation and what happens to them.**

***Methods*—A retrospective medical record study of all patients aged between 1 month and 16 years from 1983 to 1996 who required ventilation for more than 28 days.**

***Results*—Forty children were ventilated for between 36 and 180 days before discharge or death. Before their presenting illness, 13 (33%) were normal, 15 (37%) had documented predisposing conditions such as bronchopulmonary dysplasia, and the remaining 12 (30%) had diagnoses made after admission. The cause of respiratory failure was central in four patients (10%), spinal cord in eight (20%), neuromuscular in 11 (28%), and pulmonary in 17 (42%). Severe nosocomial infection requiring treatment with intravenous antibiotics occurred in 22. To date, 16 children (40%) have died, and 10 (25%) remain ventilator dependent. Of the 24 survivors, seven (29%) have severe residual neurological deficit.**

***Conclusions*—Increasingly, children are surviving intensive care only to remain ventilator dependent and at risk of significant comorbidity. This study should inform further debate on why such children remain ventilator dependent, and how and where they are managed.**

(*Arch Dis Child* 1998;**78**:253–256)

Keywords: ventilation; intensive care; outcome

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Accepted 31 October 1997

Most children in paediatric intensive care units (PICUs) with acute respiratory failure need supportive mechanical ventilation for only a short time.[1] However, there is an increasing number who require prolonged mechanical ventilation, or in whom all attempts at weaning fail so that they remain ventilator dependent.[2] Respiratory failure needing long term mechanical ventilation arises from problems with central ventilatory control, intrinsic lung disease, spinal cord damage, or neuromuscular abnormalities. Such patients inevitably have a disproportionate impact on intensive care unit resources.[3] Acute emergency admissions are restricted and prohibitive costs can arise from an inappropriately prolonged PICU stay.[4 5] It is likely that the population of ventilator dependent children will increase. Hence it is vitally important for all those involved in the provision of acute critical care to understand why some acutely ill children become ventilator dependent. This prompted us to look at our own cohort of patients over the previous 13 years to find out which children were at particular risk, the causes for their ventilator dependence, and most importantly their long term outcome.

Methods

All children aged between 1 month and 16 years admitted to the Great Ormond Street Hospital for Children PICU between 1983 and 1996 were reviewed. Those requiring positive pressure ventilation with or without a tracheostomy for more than 28 days were identified, and information was obtained from hospital notes and telephone interview. Records were reviewed for underlying pathology and whether their diagnosis was known at the time of presentation. The causes of respiratory failure were divided into the four components of the respiratory pump: central, spinal cord, neuromuscular, and pulmonary. The duration of stay was calculated. Complications of care were sought and included the need for tracheostomy or gastrostomy and the acquisition of multiresistant bacterial organisms. An arbitrary indicator of the time spent planning discharge was taken as the interval between tracheostomy and date of leaving the PICU. Outcome was analysed in terms of the need for ongoing mechanical ventilation, neurological impairment, and mortality. Neurological impairment was categorised as mild, moderate, or severe according to the following criteria: mild, delayed motor milestones and/or developmental skills; moderate, clearly retarded in motor and/or developmental skills requiring help in activities of daily living; severe, profound neurological impairment and/or mental retardation with or without an ability to interact.

Results

PATIENTS AND UNDERLYING AETIOLOGY

Between 1983 and 1996, increasing numbers of patients required prolonged mechanical ventilation (fig 1). In total 40 patients (22 boys and 18 girls), median age 4 years (interquartile range (IQR) 0.5 to 6.6 years), were mechanically ventilated for more than 28 days. None of these children had been mechanically ventilated before.

Before the presenting illness, 13 of the 40 children (33%) had no previously identified disease, 15 (37%) had an underlying identified disease, and 12 (30%) had diagnoses made after admission.

PICU STAY

The causes of respiratory failure were central in four patients, spinal cord in eight, neuromus-

Table 1 The causes of prolonged mechanical ventilation in 40 patients

Patient	Age (years)	Presentation	Underlying disease	Stay (days)	Outcome	
					Neurological impairment	Ventilator dependent
Central nervous system failure						
1	8	Cerebral sarcoid	Sarcoidosis	77	Died	–
18	3	Subdural haemorrhage	Head injury	36	Died	–
37	7	Status epilepticus	Encephalitis	104	Severe	No
38	8.9	Status epilepticus	Cerebral vasculitis	81	Severe	No
Spinal cord failure						
3	14	Transverse myelitis	Thalassaemia major	462	Severe	Yes
11	0.1	Hypoventilation	Ondine's curse	5	None	Yes
20	9.7	Cord compression	Trauma	23	Severe	Yes
22	10	Cord compression	SDT	137	Mild	Yes
23	5.2	Cord compression	SDT	98	Mild	Yes
24	5.8	Cord compression	Trauma	600	Died	–
32	5	Cord compression	Down's syndrome	730	Moderate	Yes
39	0.6	Cord compression	Anomalous spine	760	Died	–
Neuromuscular failure						
9	14	Myasthenia gravis		36	Mild	No
10	1.4	Guillain-Barré	Meningitis	112	Moderate	No
12	14	Polyneuropathy	Leukaemia	203	None	No
19	2.9	Pompe's disease		58	Died	–
21	0.4	Guillain-Barré		110	Moderate	Yes
27	1.8	Leigh's disease		189	Died	–
28	15	Leigh's disease		>910	Severe	Yes
29	0.2	Pneumonia	SMA	45	Died	–
30	6.5	Pneumonia	SMA	86	Died	–
31	1	Pneumonia	Myasthenia gravis	57	Died	–
35	7.2	Polyneuropathy	AVM	36	Mild	No
Pulmonary failure						
2	0.2	Pneumonia	Cystic fibrosis	58	None	No
4	1.3	Septic shock	Nephrotic syndrome	37	Died	–
5	0.9	Pneumonia	Cerebral palsy	41	Severe	No
6	0.1	Diaphragmatic hernia	Hypoplastic lung	180	Died	–
7	0.5	PCP	Immunodeficiency	29	None	No
8	3.7	Interstitial pneumonitis	Fanconi syndrome	35	Died	–
13	3	ARDS	Asthma	43	Died	–
14	0.9	Bronchiolitis	SCID	54	Died	–
15	0.7	Bronchiolitis	BPD	44	Moderate	No
16	0.3	Bronchiolitis	BPD	33	Mild	0_2
17	0.5	Pneumonia	BPD	57	Moderate	No
25	0.3	Bronchiolitis	OA+TOF	45	Mild	No
26	0.1	Pneumonia	CHARGE	153	Severe	No
33	0.2	Bronchiolitis	BPD	177	Mild	Yes
34	4.4	ARDS	Head injury	730	Moderate	Yes
36	0.8	Tracheobronchomalacia	Down's, AVSD, CP	385	Died	–
40	0.1	Bronchiolitis	Hypoplastic lungs	28	Died	–

Numbering is in chronological order of presentation. ARDS = acute respiratory distress syndrome; AVM = cerebral arteriovenous malformation; AVSD = atrioventricular septal defect; BPD = bronchopulmonary dysplasia; CHARGE = association of congenital anomalies; CP = cerebral palsy; O_2 = oxygen dependency; OA+TOF = oesophageal atresia and tracheo-oesophageal fistula; PCP = *Pneumocystis carinii* pneumonia; SCID = severe combined immunodeficiency; SDT = spondyloepiphyseal dysplasia tarda; SMA = spinal muscular atrophy.

cular in 11, and pulmonary in 17 (table 1). The median length of stay on PICU before death or discharge was 58 days (IQR 38 to 149 days). The median length of time from intubation until tracheostomy was 30 days (IQR 16 to 40 days), and from tracheostomy until discharge from PICU a further 78 days (IQR 44 to 254 days). Hence most chronically ventilated patients, having achieved relative medical stability, spent on average an extra 2.6 months (or nearly three quarters of the length of their hospital stay) awaiting discharge.

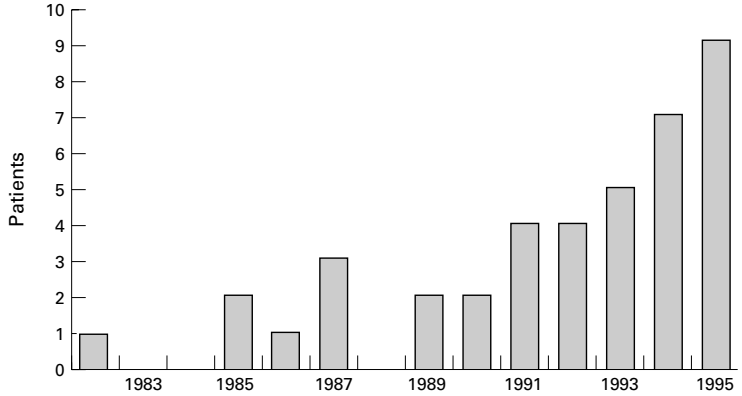

Figure 1 The number of new chronically mechanically ventilated patients seen each year (1983 to 1996).

NOSOCOMIAL COMPLICATIONS AND OUTCOME

Twenty two patients (54%) developed major nosocomial infection requiring intravenous antibiotics. Problems with methicillin resistant *Staphylococcus aureus* (MRSA) colonisation occurred in four patients and gentamicin resistant klebsiella in a further two. Nine children (22%) required a gastrostomy and 26 (65%) required a tracheostomy. Of the 40 patients, 16 (40%) have died and 10 (25%) require ongoing mechanical ventilation. Of the 16 children who died, 14 died in hospital (eight after withdrawal of mechanical ventilation, three with worsening respiratory failure, and three with sepsis syndrome), and two died at home. Of the 10 children still mechanically ventilated, two remain on our PICU (the long-

est for over 2.5 years), seven are in their local hospital, and only one is being cared for at home.

Of the 24 survivors, 20 (83%) have known neurological impairment. Of these, seven have minor impairment and are able to attend special school, six have moderate impairment, five have severe neurological impairment but retain an ability to interact, and two have severe neurological impairment with no ability to interact.

The aetiology with the worst overall prognosis was central nervous system failure, with all children dying, or surviving with severe disability and ventilator dependence. The aetiology with the next worse mortality and prognosis was neuromuscular failure, with five of 11 children dying, and a further two remaining ventilator dependent. The aetiology with worst respiratory prognosis was spinal cord failure, in which all the survivors remain ventilator dependent. The aetiology with the best respiratory prognosis was pulmonary failure, in which only two of 10 survivors remain ventilator dependent.

Discussion

Several worrying questions arise from our retrospective study. First, since 1989 the number of children requiring prolonged mechanical ventilation in our unit has increased year on year. Moreover, a significant proportion of these children on presentation have identified underlying conditions which might in some cases be regarded as inevitably lethal. Second, most of our chronically ventilated patients are remaining in hospital for lengthy periods while arrangements are made for discharge, and worryingly only one ventilator dependent child is now being cared for at home. Lastly, as a group, these children do very badly, with 42% of survivors remaining ventilator dependent and 29% having severe neurological impairment.

Our cohort of patients reflects many of the reasons why more children in the future may become candidates for long term ventilation. It is likely that we have underestimated the size of the population, given that we have concentrated on non-neonatal and non-cardiac acute disease. There is a growing population of oxygen dependent ex-premature babies with bronchopulmonary dysplasia, and there is also a greater expectation from patients with neuromuscular conditions that everything possible should be done.[6] Improved resuscitation practices and the development of specialist paediatric intensive care services has also meant that extremely sick children with acute respiratory failure now have a better chance of survival. Unfortunately, PICU physicians usually do not have control over the decision to initiate mechanical ventilation. For example, children with known neurodegenerative disorders may present at a local hospital in acute respiratory failure requiring intubation before transfer to a PICU. Only then does it become apparent that the patient is unlikely to be weaned from the ventilator, but withdrawal of support at this stage is extremely difficult.[7] Gillis and coworkers have referred to this predicament as the "phenomenon of entrapment."[8] Though acute critical illness in any child is often unexpected, in patients with known progressive conditions there should have been an opportunity to discuss the appropriateness or otherwise of ventilation. However, we accept that on occasion the futility of continued treatment only becomes clear to family members after it has been initiated.[7 9]

Key messages

- Prolonged dependence on supportive mechanical ventilation does occur as a result of acute critical illness
- Such patients are increasing in number and their problems present major medical, clinical, and ethical challenges to the paediatrician

Home care for ventilator dependent children is widely accepted as the best option for meeting the needs of child and family. However, discharge is routinely delayed for prolonged periods because of problems with funding and a lack of alternative facilities. Inevitably a proportion of these children then acquire nosocomial infections which both delays discharge further and contributes to the burden of resistant bacteria in any particular unit. Better provision should be made for regional facilities which can "offload" PICUs and provide respite care for families. In this respect, there is much we can learn from both our adult colleagues in the United Kingdom and from home care programmes in the USA, Canada, and the Antipodes, where there are well established programmes that attend to all aspects of the care of chronically ventilated children.[10]

The indications for long term ventilatory support need to be based upon good data regarding outcome. At present, uncertainty surrounding outcome compels the clinician to continue treatment despite any inclination to do otherwise. However, there are some comparative data that can guide us. In a series of 22 patients where the cause of respiratory failure was neurological in 64% and pulmonary in 31%, Canlas-Yamsuan and colleagues reported an overall mortality rate of 32%, similar to ours.[9] Not surprisingly, the reported outcome is closely related to the underlying diagnosis. Respiratory failure associated with cystic fibrosis nearly always has a poor prognosis. In 56 patients reported by Davis and di Sant Agnese, only 6% survived longer than one year.[10] On the other hand, the prognosis for children with bronchopulmonary dysplasia and tracheobronchomalacia is reasonably good. In one series, 12 of 19 children with bronchopulmonary dysplasia were successfully weaned from home mechanical ventilation.[11] It is also important that data on outcome should include an assessment of disability and quality of life. Deciding whether continuance of a seriously compromised life is "worthwhile" is always difficult and inevitably subjective. In one study of adults with quadriplegia following spinal injuries, the

majority of those questioned felt that continued mechanical ventilation was preferable to being allowed to die.[12] Similar studies need to be carried out on families of ventilator dependent children in order to try and assess how they perceive the quality of their existence. All we can say is that anecdotal evidence from such families supports the impression that many of these children do enjoy life and can develop satisfying relationships.[13]

In summary, the problems posed by chronically ventilated patients present major medical and ethical challenges to the paediatrician and intensive care specialist. A significant proportion of these children are inappropriately cared for, remain ventilator dependent, and are severely disabled. The findings of our study should inform further debate on why, how, and where such patients are managed.

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