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Clinical findings and respiratory function in infants following repair of oesophageal atresia and tracheo-oesophageal fistula

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ABSTRACT: Survival rates for infants undergoing surgical repair of oesophageal atresia with tracheo-oesophageal fistula (OA-TOF) have improved dramatically, but this condition remains associated with substantial morbidity. Most studies of patients following OA-TOF repair have concentrated on school-age and older people; whereas, the most hazardous period is infancy. We aimed to assess respiratory function in a group of infants following primary operative repair, and to relate the results to clinical findings during the first year of life.

We studied 16 infants within 3 months of primary repair of OA-TOF. Measurements were made of maximum expiratory flow at functional residual capacity ($\dot{V}$maxFRC), thoracic gas volume (TGV) and airways resistance (Raw). Ten infants had tests repeated, usually to assess progress alongside continuing symptoms, or to ascertain improvement following additional surgery.

Seven infants had essentially normal initial respiratory function tests, and six remained either symptom-free or developed only minor clinical problems. One infant subsequently developed stridor, with spontaneous improvement towards the end of the first year. The remaining nine infants had abnormal initial respiratory function tests: one was symptom-free at that time. The remainder developed respiratory and/or gastro-oesophageal symptoms. The functional abnormalities appeared to reflect the severity of the clinical problems encountered.

We conclude that respiratory function testing in infants following OA-TOF repair may augment the value of clinical appraisal, help define postoperative respiratory status, and provide a general guide to likely clinical progress.
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Until the 1940s, oesophageal atresia with tracheo-oesophageal fistula (OA-TOF) was universally fatal. Since first described in 1943 [1], primary repair has become the treatment of choice, and survival rates have improved dramatically [2]. With reduced mortality in this condition, the importance of morbidity has become increasingly relevant. In school-age children and adults, significant respiratory [3] and gastro-oesophageal sequelae [4] have been reported; but the most hazardous period for the repaired OA-TOF patient is early infancy, when increased likelihood of hospitalization [5], the occurrence of "near-death" episodes [6] and even sudden unexpected death are recognized. There have been no studies concentrating on respiratory function status in the weeks following surgery and its relationship to morbidity during infancy. The aims of our study were 1) to assess the relationship between clinical findings and respiratory function in infants following repairs of OA-TOF; and 2) to determine the value of respiratory function tests in elucidating mechanisms of respiratory disturbances and in predicting clinical outcome.

Methods

Patients

The 16 patients included in this study were born between May 1984 and February 1989. They were selected from 18 infants with oesophageal atresia and distal tracheo-oesophageal fistula presenting to the paediatric surgical service in Leicester during this period. Of the two excluded, one had multiple additional congenital abnormalities, and the other did not have lung function tests in early infancy. The group comprised 9 males and 7 females, of mean birth weight 2.71 kg (range 2.08–2.88 kg), and mean gestational age 38.6 weeks (range 35–41 weeks). Table 1 gives their clinical details, including additional diagnoses at the time of primary repair.

Study design

The clinical progress of each infant was carefully documented at out-patient visits or during admission over the

Table 1. – Clinical details at the time of repair

Patients No.	Sex	Gest. age wks	Birth weight Kg	Additional diagnoses	Gastrostomy
1	M	40	2.36	-	-
2	M	40	2.66	Bilateral hydronephrosis	-
3	M	40	3.34	-	-
4	M	39	3.10	-	+
5	M	37	2.25	-	+
6	M	38	2.22	-	-
7	M	37	2.49	Hypotonia	-
8	F	41	2.6	Anal atresia	-
9	F	39	2.61	Imperforate anus Thalassaemia trait	+
10	M	38	3.18	-	-
11	F	41	3.38	-	-
12	F	37	2.35	-	+
13	F	37	2.43	-	-
14	F	38	3.10	-	+
15	M	40	2.78	-	-
16	F	35	2.08	-	+

Gest: gestational; M: male; F: female.

first year, with particular reference to feeding difficulties, including choking, respiratory symptoms including cough or stridor, and the occurrence of "near-death" episodes. Measurements of respiratory function were made soon after repair, and the nature and severity of abnormalities were related to the clinical status of each child.

Nine infants had their initial respiratory function tests within a month of surgery, and all were studied within 3 months. Repeat testing was carried out in 10 infants (cases Nos 1, 2, 8 and 10–16), in 8 cases because of persistent or recurrent clinical problems following surgery. Four infants were studied on three occasions: 3 as a sequel to tracheopexy or aortopexy (cases Nos 13, 14 and 16), and one following recurrent cyanotic episodes attributed to aspiration (case No. 8). Informed consent for lung function tests was obtained from the parents of all infants, and parents were usually present throughout the tests. On each occasion, when an infant was retested, the results were assessed in relation to clinical findings at that time.

Measurements of respiratory function

Assessment of respiratory function consisted of plethysmographic determination of thoracic gas volume (TGV) and airways resistance (Raw) [7], and measurement of maximal respiratory flow at function residual capacity ($\dot{V}$maxFRC) using the squeeze technique [8]. Infants were sedated with chloral hydrate (100 mg·kg^{-1} body weight), and once asleep were wrapped in an inflatable jacket (RPMS Workshops, Hammersmith Hospital, London, UK) extending from the neck to the thighs. A facemask with a screen pneumotachograph (Erich Jaeger, GmbH) attached was applied around the nose and mouth, using a ring of sterile putty to form an airtight seal. Respiratory flow was electronically integrated to provide a tidal volume signal, and the flow-volume loop was displayed on an oscilloscope (Telequipement Type D1011). At the end of a tidal inspiration, the jacket was rapidly inflated by opening a three-way tap connected to an air-filled pressure reservoir. The resultant compression of the chest and abdomen caused a rapid expiration, producing a partial forced expiratory flow-volume curve. Signals of respiratory flow, volume, and jacket pressure were recorded on a storage oscilloscope (Bodytest, Erich Jaeger GmbH) and subsequently plotted at 1/4 speed onto a chart recorder. These signals were also stored on an Apple IIe microcomputer for later analysis. Between 8–12 partial forced expiratory flow-volume curves were recorded for each infant, over a range of applied jacket pressures up to a maximum of 80 cmH_2O (7.84 kPa). A period of approximately 40 s was allowed between chest compression, to allow tidal volume and end-expiratory level to stabilize. After 3 or 4 consecutive measurements, the facemask was removed for 1–2 min as recordings were inspected.

Recordings were inspected and only those technically acceptable were analysed [9]. The final recorded value of $\dot{V}$maxFRC was the highest value obtained from the flow-volume loops analysed.

When the squeezes were completed, the jacket was loosened and the infant continued to sleep within the whole-body plethysmograph. Thoracic gas volume was measured according to the method of DUBOIS *et al*. [10], as modified for infants [11]. A new facemask, with an additional outlet to measure mouth pressure, was applied and attached to a rebreathing apparatus *via* two pneumatically operated valves, allowing the infant to breathe from either the plethysmograph or a heated rebreathing bag. After a brief period breathing warmed air, usually well within a minute, both valves were closed for a few seconds to produce total airway occlusion against which the infant made several respiratory efforts. The recorded signals of flow, plethysmographic and mouth pressures were stored both in the oscilloscope and on computer, as above, and used to calculate TGV and Raw.

Analysis of respiratory function tests

Forced expiratory flow-volume curves were visually inspected and each subject was classified as having either a variable, irregular pattern of flow or a more smooth pattern (fig. 1). This classification was carried out by one person (CSB), and was performed by eye. In subjects with a smooth pattern, V̇maxFRC was measured [12] and expressed as number of standard errors of prediction away from the predicted value for that infant. In patients with an irregular pattern of flow, no reliable value of V̇maxFRC could be calculated.

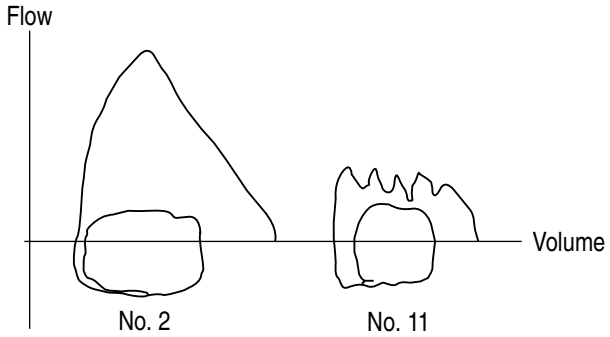

Fig. 1. – Patterns of flow-volume curves: left, infant No. 2, normal pattern; right, infant No. 11, irregular pattern.

Measurements of TGV were used to determine whether infants had a small or a normal lung volume, in which case TGV was within or below 2 SE of the predicted value; or if they were hyperinflated, defined as TGV greater than predicted value +2 SE.

Airways resistance was measured from the recordings of flow and pressure taken from the period of quiet breathing before the airway occlusion for TGV measurement. At least five representative breaths were chosen from the charted and computerized records to calculate airways resistance over 2/3 initial inspiratory flow.

For all three measurements (V̇maxFRC, TGV and Raw) predicted values and standard errors of prediction were calculated from data obtained from normal infants studied within the first year of life [13].

Patterns of airways resistance were also categorized according to pressure-flow loops: 1=normal; 2=abnormalities predominantly during expiration; 3=abnormalities during inspiration; 4=abnormalities during both phases of the respiratory cycle (fig. 2). This classification was performed by eye, by one investigator (CSB).

Limitation of the methods

Although respiratory function testing would ideally have been carried out in all infants at predetermined ages, the number and timing of tests was of necessity determined in part by: 1) the clinical status of each individual infant; and 2) the willingness and ability of parents to return for infant testing. Several infants lived outside Leicester and its immediate surroundings, and some parents were unwilling to return for infant tests which could be protracted and which required sedation.

Similarly, clinical assessments were carried out at scheduled appointments in out-patients clinics for all infants, but those infants who were admitted to the hospital or who attended for additional investigation were seen more often that those who remained well. Thus, close temporal matching of clinical and respiratory function data was not always possible for all infants.

Investigations other than respiratory function tests were carried out in infants only when clinically relevant. For example, it would not be considered justifiable to undertake investigations of possible gastro-oesophageal reflux in infants who have no gastrointestinal symptoms. Thus, there was considerable variation in the extent and type of investigation within the study population.

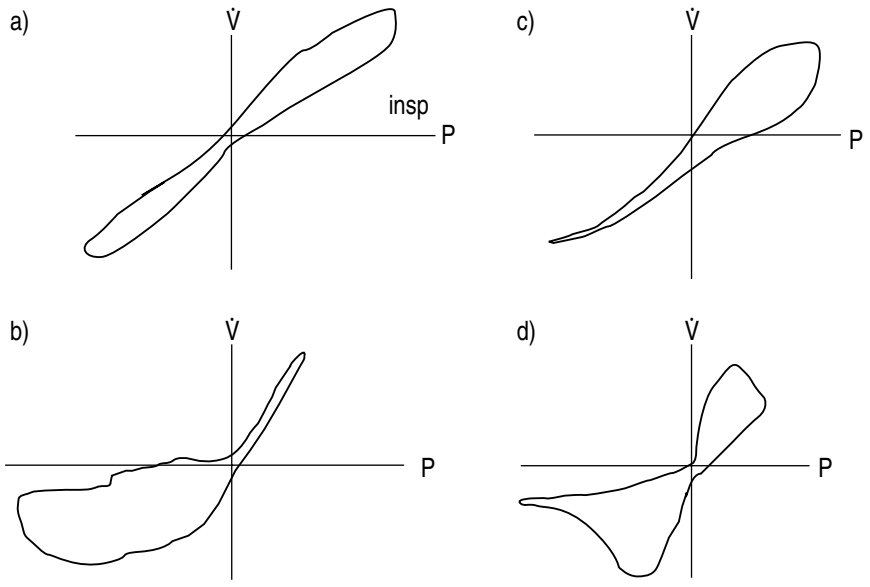

Fig. 2. – Patterns of pressure-flow loops. a) Infant No. 11, normal pressure-flow pattern, type 1. b) Infant No. 13, distortion of expiratory portion of pressure-flow pattern, type 2. c) Infant No. 12, pressure-flow loop shows distortion during inspiration, pattern type 3. d) Infant No. 7, pressure-flow loop distorted during inspiration and expiration, pattern type 4. V̇: flow; P: pressure; insp: inspiration; exp: expiration.

The analysis of shape of the pressure-flow loops and flow-volume curves was performed by eye, and was, therefore, subjective. However, all curves were analysed by one person. At the time of the initial investigation she had no knowledge of the site or extent of repair, and obviously could not predict the clinical outcome. The person analysing the respiratory patterns played no part in clinical evaluations. At the time of subsequent respiratory function tests she could not, for practical reasons, be completely blind to the clinical course of any individual, but she was unaware of the results of any additional investigations an infant might have undergone.

Results

Initial findings

Table 2 summarizes the results of respiratory function tests. Table 3 indicated the main clinical problems encountered prior to the initial tests, together with measurement of respiratory function expressed as standard error scores. Infants Nos. 1–5 were symptom-free and showed no abnormalities of lung function, other than low airways resistance (two patients). Infants Nos. 6–8 had relatively mild antecedent symptoms at this stage, with case No. 7 showing abnormal lung function. This infant had an earlier cyanotic episode unrelated to feeding and had been noted to be hypotonic. He demonstrated various patterns of inspiratory and expiratory airways resistance and irregularities of flow-volume curve. The results were consistent with limitation of inspiratory and expiratory airflow (fig. 2). The abnormalities observed were perhaps related to recent surgery, hypotonia, and need for a chest drain within days of testing.

The remaining infants (cases Nos. 9–16) all had significant antecedent symptoms and abnormalities of lung function when first tested. All were clinically stable at that time. Symptoms were predominantly respiratory or gastro-oesophageal, and lung-function abnormalities included an increase in TGV (cases Nos. 11, 14, 15), abnormalities in pattern of Raw (cases Nos. 9, 10 and 12–16), and increases in Raw (cases Nos. 10, 14 and 15). Precise interpretation of these results in relation to the symptoms observed is not always possible, but in most cases it is consistent with significant upper and/or lower respiratory pathology.

Progress during the first year of life

Table 4 gives the clinical and lung function findings for 10 cases, for whom follow-up lung function data are available.

Infants who were initially symptom-free with normal lung function had uneventful courses during the first year.

Infants Nos 1–5 remained symptom-free. Tests were repeated in infants Nos 1 and 2 because of parental anxiety, but results were essentially normal. Infant No. 6 developed soft inspiratory stridor and progressed normally. Infant No. 7 had occasional "noisy" breathing. Follow-up was not possible for this case, but postal questionnaire suggested that he had no further symptoms. Case No. 8 developed stridor by the age of 7 weeks, and was admitted to hospital at 16 weeks following an apparently life-threatening episode that occurred during feeding. Investigations revealed swallowing inco-ordination, stricture of the oesophagus, and gastro-oesophageal reflux. The lung function abnormalities noted were consistent with tracheal instability and tracheobronchial aspiration. Symptoms resolved later in the first year and tests at 46 weeks were normal. Case No. 8 was the only infant with relatively minor symptoms and normal lung function initially who developed significant clinical problems subsequently. These could be attributed to upper and lower respiratory tract complications.

Infants Nos 9–16 remained symptomatic and, except for case No. 12, repeat lung function tests remained

Table 2. – Pulmonary function tests - raw data

Pt No.	Age at test wks	Length cms	TGV ml	Raw $cmH_2O \cdot l^{-1} \cdot s$	$\dot{V}maxFRC$ $ml \cdot s^{-1}$
1	3	47.8	75	P	82
	38	67.5	167	10.7	147
2	2	50.7	77	P	148
	33	67.7	174	P	329
3	6	50.5	94	26.2	131
4	2	47.5	92	19.7	59
5	2	47.8	107	16.0	88
6	3	46.0	75	30.7	-
7	2	45.5	91	53.9	IR
8	4	50.2	79	P	124
	16	60.5	156	46.1	IR
	46	72.2	182	14.2	97
9	6	52.3	89	37.0	32
10	8	60.7	199	93.5	-
	51	80.8	312	21.1	189
11	12	58.6	108	20.7	IR
	45	65.8	173	28.6	IR
12	6	48.9	101	45.7	35
	18	53.8	121	28.9	55
13	13	53.0	112	36.0	-
	32	61.2	219	17.0	-
	45	64.8	196	24.0	255
14	9	56.5	177	46.3	-
	21	59.0	238	30.0	-
	37	65.7	264	24.8	-
15	3	48.0	129	93.9	-
	10	55.3	142	14.0	47
16	2	46.3	108	64.1	-
	9	50.5	169	53.0	79
	13	55.2	124	64.7	85

Pt: patients; TGV: thoracic gas volume; Raw: airways resistance; P: pattern only; $\dot{V}maxFRC$: maximum expiratory flow at functional residual capacity; IR: variable irregular pattern - no reliable value could be assigned.

Table 3. – Results of initial respiratory function tests and concurrent clinical findings

Pt No.	Age at Test 1 wks	Pretest symptoms	TGV (SE score)	Raw pattern	Raw (SE score)	V̇maxFRC (SE score)	Comment and interpretation
1	3	-	-0.90	1	P	+0.12	Normal results consistent with absence of symptoms
2	2	-	-1.74	1	P	+1.10	
3	6	-	-0.48	1	-0.36	+0.80	
4	2	-	+0.31	1	-2.52	-0.32	
5	2	-	+1.29	1	-4.00	+0.24	No reason for low value of Raw; no symptoms
6	3	Occasional harsh cough	-0.28	1	-0.59	-	Normal results; minimal symptoms
7	2	Cyanotic episode unrelated to feed; hypotonia	-0.03	4	+0.08v	IR	See text
8	4	Stridulous cry at 2 weeks	-0.52	1	p	+0.66	Normal results not suggestive of extrathoracic narrowing at this time
9	6	Cough with feeds; persistent chest signs	-1.43	2	+1.42	-1.29	Results consistent with lower respiratory involvement
10	8	Preoperative pneumonia; choking during previous week	+0.43	2	+5.30v	-	Results consistent with tracheo-bronchial aspiration; endoscopy normal
11	12	Feeding difficulties; choking episodes and recurrent vomiting	+2.90	1	+0.04	IR	Results consistent with recurrent aspiration pneumonia. Oesophageal stenosis and GOR confirmed; endoscopy at 6 months showed a second fistula
12	6	Convulsions on day 2; Inspiratory stridor	+0.53	3	+1.47	-0.89	Results consistent with extrathoracic narrowing; subglottic stenosis confirmed by endoscopy
13	13	Recurrent aspiration pneumonia; stridor	-0.18	2	+1.47	-	Findings consistent with lower respiratory involvement; GOR and tracheomalacia confirmed
14	9	Biphasic stridor; feeding difficulties; cyanotic and "near-death" episodes; oesophageal stenosis	+2.16	4	+3.32	-	Inspiratory and expiratory abnormalities due to tracheomalacia and tracheobronchial aspiration
15	3	Choking with feeds; persistent chest signs; stridor absent	+2.54	3	+2.60v	IR	Results suggestive of extrathoracic obstruction and lower respiratory involvement; lack of stridor was surprising
16	2	Periods of mechanical ventilation for 8 days post-operatively; respiratory distress on extubation	+1.86	4	+1.62v	-	Results consistent with tracheomalacia - confirmed endoscopically

Thoracic gas volume (TGV): numbers shown are standard errors (SE) of prediction airway from mean predicted value (values between -2 and+2 are considered normal). Airways resistance (Raw): 1=normal pattern and 2–4=abnormal patterns (see fig. 2); numbers shown are standard errors of prediction; P:pattern only; v=marked breath-to-breath variability. V̇maxFRC: flow volume curve; -=measurement not made; numbers shown are standard errors of prediction for V̇maxFRC of curves which could be analysed. GOR: gastro-oesophageal reflux. For further abbreviation see legend to table 2.

abnormal. Feeding difficulties ranged from slowness to feed, vomiting, choking and suspected aspiration. Gastro-oesophageal reflux was confirmed in five infants (table 4), and two (cases Nos 11 and 13) were treated surgically by Nissen's fundoplication. Three infants (cases Nos 11, 12 and 14) developed airway narrowing or stricture of the oesophagus, for which dilatations were necessary. Clinical signs of lower respiratory involvement were common, and attributed either to tracheo-bronchoaspiration or intercurrent chest infection. Upper airway problems included mild to severe stridor, "near-death" episodes attributed either to tracheal collapse or choking, with suspected aspiration. Among infants with stridor, patterns of airways resistance characteristic of intrathoracic upper airway obstruction were observed in 3 of 6 infants. These were reminiscent of patterns seen

Table 4. – Results of later respiratory function tests and concurrent clinical findings

Pt No.	Age at Test wks	Stridor	"near-death" episodes	GI symptoms*	GOR	TGV (SE score)	Raw pattern	Raw score	$\dot{V}$maxFRC (SE score)	Comment and interpretation
1	38	-	-	-	-	+2.09	1	-3.88	-0.60	No serious abnormalities of pulmonary function. Both infants remained very well
2	33	-	-	-	-	-1.88	1	P	+2.88	
8	16	+	+	+	+	-0.26	3	+3.85	IR	Results consistent with extrathoracic airway narrowing, exacerbated by choking and GOR
	46	-	-	-	-	-1.92	1	-0.06	-1.99	Normal respiratory function tests coincide with clinical recovery
10	51	-	+	+	+	-0.36	1	+4.52	-1.07	Results remain consistent with aspiration
11	45	+	-	+	+	+3.16	1	+3.26	IR	GOR treated surgically; results consistent with continuing lower respiratory involvement
12	18	+	-	+	-	+0.09	1	+0.77	-0.99	Normal results despite symptoms
13	32	+	+	+	+	+2.50	2	-0.76	-	Similar to first test; hyperinflation consistent with continuing aspiration
	45	-	-	+	+	+0.23	1	+2.24	+1.75	Improvement following tracheopexy at 40 weeks; residual increase in airways resistance consistent with continued aspiration; surgical treatment of GOR subsequently
14	21	+	+	-	+	+4.07	4	+2.04	-	No change since first test
	37	-	-	+	+	+2.82	1	+2.52	-	Improved since tracheopexy at 34 weeks; persisting hyperinflation and raised resistance
15	10	+	+	+	+	+0.77	1	-3.62	-1.30	Improved lung function despite persistent symptoms
16	9	+	+	+	-	+3.82	4	+2.08v	-0.20	Abnormalities detected are consistent with upper and lower airway problems
	13	+	+	+	-	-0.21	4	+3.53v	-0.56	Pattern of airways resistance not improved by aortopexy at 10 weeks

GI symptoms: gastrointestinal symptoms include feeding difficulties, choking and suspected aspiration. For abbreviations see legend to table 2 and 3.

in infants with tracheal vascular rings [14], and were compatible with intrathoracic tracheomalacia.

Three infants underwent tracheopexy (cases Nos 13, 14 and 16), with immediate clinical improvement in two. Infant No. 16 also had aortopexy but did not improve clinically following either operation, and subsequently died. Case No. 15 developed symptoms related to upper airway obstruction, gastro-oesophageal reflux and suspected aspiration. Despite this, his lung function tests had much improved when repeated at 10 weeks. He died suddenly at 20 weeks, after an illness initiated by a choking attack. Klebsiella septicaemia and necrotizing enteritis were diagnosed at postmortem examination. The site of repair of the tracheo-oesophageal fistula was intact, and there was no occlusion or narrowing of the air passages.

Case No. 16 had considerable feeding and respiratory difficulties throughout her life, due to extensive tracheomalacia. The operative procedures carried out did not lead to clinical or function improvement. She died at 15 months of age with a chest infection, and permission for postmortem was not obtained.

Only two infants in the series (cases Nos 9 and 10) had persistent symptoms attributable only to gastro-oesophageal complications. Both had initial functional abnormalities consistent with aspiration; repeat testing of case No. 10 showed little change. The symptoms and lung function results seen in case No. 11 were consistent with recurrent/chronic aspiration, confirmed with discovery of a second fistula at 6 months of age. Lung function tests following Nissen's fundoplication and repair of fistula were still consistent with lower respiratory involvement towards the end of the first year.

Follow-up respiratory function tests showed persisting abnormalities in cases Nos 8 and 10–16. The relative contributions of upper and lower respiratory pathology to the functional abnormalities detected were not easy to separate (table 4). Unfortunately, endoscopy was not carried out in all infants with stridor, although tracheomalacia was confirmed by this means in those treated by

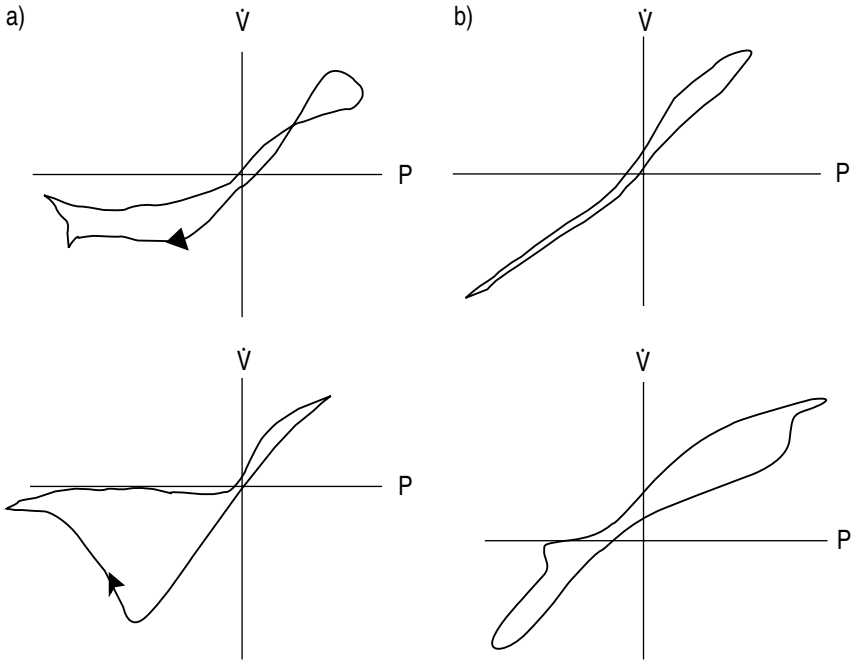

Fig. 3. – Pressure-flow loops from infants Nos 13 and 14. a) Patterns before additional surgery. b) Patterns following tracheopexy. V̇: flow; P: pressure.

tracheopexy/aortopexy (cases Nos 13, 14 and 16). When upper respiratory symptoms (predominantly stridor) dominated, patterns of respiratory resistance were frequently abnormal. Choking and "near-death" episodes can result from upper airway obstruction or tracheobronchial aspiration. Infants with these symptoms all had abnormalities of lung function, but whether these were primary determinants of symptoms or secondary to them was not always clear from the history or the pattern of abnormalities detected.

Two infants (cases Nos 13 and 14) showed a reduction in hyperinflation and improvement in pattern of airways resistance following tracheopexy. Figure 3 illustrates pattern found before and after surgery.

Discussion

These results present an opportunity to comment on respiratory function in infancy following OA-TOF repair, and to correlate functional indices of severity with clinical findings and complications during the first year of life. We have demonstrated abnormalities in pulmonary function in 9 of 16 (56%) infants investigated up to three months following OA-TOF repair. The nature and extent of these abnormalities broadly correspond with the occurrence of symptoms reported at that time.

In general, the more severe the functional abnormalities on initial testing the greater the range and severity of persisting symptoms. The extent to which symptoms are determined by, or subsequently accounted for, lung function abnormalities could not always be assessed. Many infants developed both upper respiratory and gastro-oesophageal complications each a potential cause of respiratory dysfunction. It is not surprising, therefore, that the relative contribution of these complications to the functional abnormalities detected (or *vice versa*) was not always clear. Respiratory symptoms *per se* seldom indicated a particular pattern or dysfunction. These symptoms dominated in the most severely affected infants in the series, and among six with "near-death" episodes five had inspiratory and/or expiratory stridor, five proven gastro-oesophageal reflux, and two had had repeated dilatations for oesophageal stricture. It is tempting to speculate that functional abnormalities in the upper airway accounted for the upper respiratory tract symptoms reported, and that tracheobroncho-aspiration was the main cause of lower respiratory symptoms and dysfunction. Whether or not the functional abnormalities detected were of primary or secondary importance, the results clearly indicate that symptomatic infants with lung function abnormalities in the week following operation merit close surveillance throughout infancy and, when indicated, active treatment for complications that arise.

There have been no previous studies relating clinical and respiratory function findings during the first year following OA-TOF repair. Studies in older children provide a different perspective. Persistence of respiratory symptoms and the extent of functional abnormalities is variable. MILLIGAN and LEVISON [3] studied lung function following OA-TOF repair in children aged 7–18 yrs and found that all but one of 24 children followed-up had abnormal respiratory function. There was no relationship between the nature and degree of abnormality demonstrated and clinical course during infancy. In a later controlled study of 20 children, aged 8–17 yrs, with a specific abnormality (Vogt-type III B), COURIEL *et al.* [15] showed that mild restrictive defects were common, and that the flow-volume loops of these patients were indicative of proximal obstruction, which was, at least in part, extrathoracic in location. Eighteen of the 20 subjects had recurrent bronchitis, and 17 had been admitted

to hospital at some point with pneumonia. There was a significant correlation between the extent of respiratory problems and oesophageal problems.

In one recent large study, respiratory morbidity was assessed in 334 patients, aged 1–37 years, showing that almost half the group needed readmission to hospital with respiratory illness, usually within the first 5 yrs of life [5]. Low birth weight patients and those with gastro-oesophageal reflux (GOR) were more likely to be admitted. Episodes of airway obstruction which were unrelated to feeding and which resulted in cyanosis were described in 8% of patients aged 0–5 yrs but were not present in older patients.

In a group of 18 patients, aged 12–21 yrs, with the commonest basic defect (Vogt-type III B) a history of pneumonia before the age of 4 yrs, was associated with reduction in forced vital capacity (FVC), forced expiratory volume in one second (FEV_1) and total lung capacity (TLC) [16]. One aim of that study was to determine whether GOR secondary to lower oesophageal sphincter incompetence was responsible for respiratory problems. The presence or absence of GOR was found to be unrelated to respiratory function status, but abnormal oesophageal motility was found in all patients. This was proposed as an alternative cause of aspiration.

A study of 12 adult patients [4] found that they were largely asymptomatic from a respiratory standpoint, but had mild restrictive lung disease without evidence of airflow obstruction. Eleven had some dysphagia and 9 suffered from heartburn. Reflux was common and several patients had structural oesophageal abnormalities. All showed lack of peristalsis in at least one portion of the oesophagus. In contrast to the study of COURIEL *et al*. [15], where a correlation between oesophageal symptoms and respiratory problems was found, there was no correlation between pathological GOR and spirometry results. There was, however, considerable scope for selection bias, as only 12 of 50 patients invited to participate responded.

These studies in older children and adults place more emphasis on the respiratory complications of gastro-oesophageal dysfunction than on those related to central airways obstruction due, for example, to tracheomalacia. Improvement with age may explain the diminishing impact of this structural abnormality. In the present study, during infancy the relationship between upper airway obstructive episodes, GOR and aspiration were not always clear; morbidity is greatest when all of these problems were demonstrated in the same individual. Effective anti-reflux treatment, or surgical relief of upper airways obstruction, may prove effective in reducing symptoms and improving lung function, as in cases Nos 13 and 14. Case No. 15, who died suddenly despite satisfactory respiratory function tests at 10 weeks of age, was awaiting investigation for gastro-oesophageal reflux. His terminal illness started with vomiting and subsequent diarrhoea, suggesting a primary infective aetiology or oesophageal dysmotility-related aspiration.

Aortopexy has been show to have a place in the treatment of infants with tracheomalacia associated with OA-TOF repair [6, 17]. However, in patients with primary tracheomalacia, it has been shown to be unsuccessful in those patients with coexisting GOR [18]. These authors strongly recommend aggressive anti-reflux .treatment for patients with these problem. This approach, also proposed for infants following TOF repair [3] concurs with our own experience. Respiratory function testing of infants following OA-TOF repair is now part of our routine clinical assessment. An argument can also be made for more aggressive assessment of oesophageal function, but in the present study this was limited to infants with significant feeding and nutritional problems.

In conclusion, primary repair of OA-TOF can be surgically successful, whilst leaving functional abnormalities of the oesophagus and trachea [19]. In patients surviving into adult life, such abnormalities may cause little or no residual handicap [20], whereas in infancy they may cause life-threatening illnesses [21]. Our experience bears this out, and provides new information on lung function in such cases. Clearly, it is essential to detect those at risk of the most serious problems as early as possible. The investigations that we have described go some way towards identifying the functional consequences of postrepair abnormalities, but, more importantly, offer the possibility of assessing any improvement resulting from further medical or surgical treatment or indeed from growth and development. Our study suggests that it is probably unnecessary to subject every infant to repeated investigations to define lung function and GOR status, but in those with signs of upper airways compromise, particularly of an episodic nature, comprehensive evaluation of both pulmonary and oesophageal function can be helpful in establishing pretreatment status and the effects of any intervention.

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