ORIGINAL ARTICLE

Home Oxygen Therapy for Children with Chronic Lung Diseases

M Z Norzila, MMed*, B H O Azizi, FRCP, A W Norrashidah, MMed, N M Yeoh, SRN, C T Deng, MMed, *Jabatan Pediatrik, Institut Pediatrik, Hospital Kuala Lumpur, **Damansara Specialist Hospital, ***Subang Jaya Medical Centre**

Summary

Home oxygen therapy programme is new in Malaysia. This programme enables children with respiratory insufficiency to be discharged home early.

Materials and Methods: Long term oxygen therapy was initiated using an oxygen concentrator in patients who i) remained hypoxic while breathing room air, ii) experienced desaturations of more than 20% during sleep as seen in patients with severe laryngomalacia and obstructive sleep apnoea syndrome and iii) had pulmonary hypertension with or without polycythaemia. The median with first and third quartile values are presented for the quantitative variables.

Results: A total of 71 patients mainly children with bronchopulmonary dysplasia (BPD) (32) and bronchiolitis obliterans (12) were discharged home on this programme. The median age at which home oxygen was initiated in children with BPD was 5.0 (Q1: 2, Q3: 8) months. The median total duration of oxygen requirement for BPD was 8.0 (Q1: 5, Q3: 12) months. The median duration of home oxygen dependency was 3.5 (Q1: 3, Q3: 6) months. However children with bronchiolitis obliterans required longer duration of oxygen therapy compared to children with BPD i.e. median duration of 28 months (Q1: 14.5, Q3: 66). In other respiratory conditions the mean duration of supplemental oxygen varies some of which may be life long.

Conclusions: This paper has shown the importance of home oxygen program in children with respiratory disorders. It has significantly shortened hospital stay and thus saves hospital costs and prevents prolonged separation from the family.

Key Words: Respiratory insufficiency, Home oxygen therapy, Bronchopulmonary dysplasia, Bronchiolitis obliterans

Introduction

Children with respiratory insufficiency requiring long term oxygen therapy or ventilatory assistance constitute of a heterogeneous population. Paediatric conditions leading to chronic respiratory insufficiency include BPD, interstitial lung disease, bronchiolitis obliterans, cystic fibrosis, neuromuscular disorders that restrict chest wall movement, obstructive sleep apnoea, central

hypoventilation syndrome and other congenital lung disorders[1]. These patients may become oxygen dependent and require supplemental oxygen in order to be discharged home.

The development of new technology has made it possible to establish home oxygen management programmes in paediatric respiratory centres. These programmes have allowed early discharge of patients and more importantly, have improved patient's quality of life both physically and emotionally[2,3].

In Malaysia the paediatric respiratory home oxygen program was started in 1992 by Azizi in Universiti Kebangsaan Malaysia (UKM) for a group of respiratory patients who were oxygen dependent[4]. The first patient who was discharged home was a case of Wilson Mikity syndrome who had to stay for a year in hospital because of oxygen dependency. She was finally discharged home using an oxygen concentrator to deliver the required supplemental oxygen.

A total of 12 oxygen concentrators donated to the unit were made available to patients. These concentrators were loaned to patients who lived in the Klang Valley on short term duration of about six months to one year. Patients who lived outside the Klang valley and who were referred from the other states such as from Pahang, Sabah or Sarawak and needed oxygen therapy permanently, were required to purchase a concentrator.

Since the commencement of this programme a total of 71 cases were on home oxygen therapy. This paper reports our experience with home long-term oxygen therapy among paediatric patients in Malaysia.

Materials and Methods

Initiation and management of home oxygen therapy

Long term oxygen therapy was initiated in i) patients who remained hypoxaemic while breathing room air, ii) patients with desaturation of more than 20% during sleep as seen in patients with severe laryngomalacia and obstructive sleep apnoea syndrome and iii) patients with pulmonary hypertension with or without polycythaemia.

Discharge policy for children on home oxygen therapy

The patients were considered for discharge when they had no more active medical problems, were feeding well and the need for supplemental oxygen was the only reason they remained in hospital.

There should be no more episodes of frequent severe bronchospasm in children who exhibited bronchial hyperreactivity. Their bronchodilator requirements had been reduced to at least four to six hourly and may be delivered through a metered dose inhaler via a spacer with a face mask.

A trained nurse (NMY) would make a home visit. During the visit the patients' house was assessed in terms of locality, surrounding environment and fire hazards. Ideally the house should be a brick house with a room identified to place the concentrator. The room should not be too near the kitchen and away from an area where there is burning of rubbish. There should be no smokers in the house. If the patient's house was a wooden house or they lived in an apartment (above the first floor), they would not be recommended for home oxygen therapy to avoid any fire hazards to the family and surrounding neighbours. Parents were recommended to make appropriate adjustments and changes to the house to fulfill the standard of safety. If the safety requirements were not fulfilled, the patients would not be sent home. Arrangements would then be organised to send them to the nearest hospital nearest their home.

Oxygen concentrator

The equipment used for home oxygen therapy was an oxygen concentrator which concentrates oxygen from the air. This electrically operated

machine draws in room air through a molecular sieve, which separates oxygen from the other gases and deliver oxygen to the patient in the required amount. It delivers oxygen in the range of 0.2 l/min to 5 liter/min via a nasal cannula.

The concentrator was placed in an identified room in the house recommended by the visiting nurse. It should not be moved from one room to another or transported from one address to another as the machine may be easily damaged. In order to facilitate transportation to the hospital for clinic follow-up, in situation when the electrical supply to the patient's home was interrupted or the concentrator was faulty, a four-liter portable oxygen cylinder was required as a back up during these situations.

Parental education

Parents were adequately informed of the disease of the patient. They were taught how to implement all safety procedures when patients were on home oxygen therapy. They were taught to recognise symptoms of early respiratory distress in their child and to seek medical attention early. They were taught proper use of medications particularly the metered dose inhalers.

Regarding the care of the concentrator, parents were taught to clean the sponge filter every alternate day and to change the humidifier water and clean the tubing with soap and water every day. The working of the concentrator, its basic operations and functions and detection of any warnings of a breakdown of the machine were taught to parents. In an event of a breakdown, the servicing company involved would be immediately contacted.

Prior to discharge the patient would be on the concentrator for at least 24 hours in the ward and vital signs i.e. oxygen saturation, respiratory rate and heart rate were monitored. If these parameters were satisfactory, the patient was discharged.

Monitoring of patients

After discharge, the patients were seen in two weeks in the Day Care Unit in Paediatric Institute. They did not attend the usual specialist clinic to minimize infections by avoiding hospital crowds in the waiting area and waiting time. They were connected to the hospital's oxygen source to minimize the use of their portable oxygen. During the review, the patient's growth parameters and oxygen saturation would be measured using a Pulse oximeter (Nellcor). A dietitian was also involved in the care of these patients. Adequate calories were required to maintain normal growth Their caloric requirements are usually higher (10 - 20% more) compared to normal children because of their increased work of breathing. In addition, their vaccination schedules were updated. Many of them particularly, patients with bronchopulmonary dysplasia had delayed vaccination due to prolonged hospitalisation, intercurrent infections and treatments with oral steroids. They were also given direct access to the respiratory ward when they were sick. This was to avoid long waiting time in the Casualty Department and being seen by medical personnels who were not well versed with their problems. Once they were stable and showed improvement, they would be seen monthly.

Discontinuation of oxygen

When they were able to maintain arterial oxygen saturation at 94% or above in air during the day, the oxygen supplementation would be discontinued in the day. The parents were then advised to recommence day oxygen if they had difficulty feeding or they developed upper respiratory tract infections.

Prior to discontinuing nighttime oxygen the patients would be admitted for an overnight oximetry. Nighttime oxygen was discontinued if the patients were able to maintain a Sa02 at 94% or higher.

Statistics

Results were tabulated and results were expressed as median and interquatile range. Analysis was performed using the Minitab Version 3.

Results

Clinical characteristics of patients

During the seven year period (1992- Dec 2000), a total of 71 cases were sent home on this programme. (Table I)

There were 50 males and 21 females. The ethnic distributions were, 47 Malays, 20 Chinese, three Indians and one Sabahan.

The majority of patients had (BPD). The mean gestational age for these patients was 28 weeks. The median age of initiating home oxygen therapy was 5 (Q1:2, Q3:8) months. The duration of home oxygen dependency was 3.5 (Q1: 3, Q3: 6) months. The total duration of oxygen requirement for BPD was 8.0 (Q1: 5, Q3: 12) months. Two patients died from severe bronchopneumonia in the first year of life. Eight patients had gastro-oesophageal reflux and six were fundoplicated. Twenty-two patients suffered from hyperactive airway disease and required long-term inhaled steroids and intermittent bronchodilators.

The second most common respiratory problem that required home oxygen therapy was bronchiolitis obliterans. Bronchiolitis obliterans is a disease of the respiratory bronchioles and alveolar ducts characterized by luminal plugging with granulation tissue or luminal obliteration by fibrosis. It commonly follows respiratory tract infections due to adenovirus, influenza virus, measles or Mycoplasma *pneumoniae*[5]. It may also be caused by recurrent aspirations from underlying gastro-oesophageal reflux[6]. The damage to the lung may be severe and may result in oxygen dependency; without oxygen supplementation, cor pulmonale may ensue.

All our patients with bronchiolitis obliteran were males. The median age of commencement on oxygen concentrator was 17 months (Q1:6, Q3:20 months). As to the causes, five patients had severe gastro-oesophageal reflux. Three of them

Table I
Types of Cases and Duration of Oxygen Dependency

Diagnosis	Numbers	Median age (months) on initiation of oxygen concentrator therapy	Median duration (months) of oxygen dependency	Median duration (months) on home oxygen therapy	No of deaths
Bronchopulmonary dysplasia	32	5 (Q1:2 Q3:8)	8 (Q1:5, Q3:12)	3.5 (Q1:3, Q3:6)	2
Bronchiolitis Obliterans	12	17 (Q1:6, Q3:20)	28 (Q1:14.5, Q3:66)	19 (Q1:1, Q3:48)	1
Congenital abnormalities	11	5 (Q1:2.5, Q3:7)	6 (Q1:4, Q3:9.5)	3 (Q1:0.75, Q3:5.5)	6
Interstitial lung disease	4	6.5 (Q1:3.5 Q3:8)	10 (Q1:6.8, Q3:53.7)	3 (Q1:3, Q3:60)	0
Cardiac diseases	5	6.5 (Q1:4.3 Q3:8.8)	4	9	3
Bronchiectasis	3	5	10	5	1
Wilson Mikity Syndrome	2	4, 9	5	1	
Obstructive Sleep Apnoea	1	1	still on (7years)	still on (7 years)	0
Intraspinal Haemorrhage	1	12	-	6	1

Q1: First Quartile; Q3: Third Quartile

Table II
Causes of Death in Patients that were on Home Oxygen Therapy

Diagnosis	Age at death (months)	Cause of death
Jeune asphyxiating thoracic dystrophy	9	Severe pneumonia
Bronchopulmonary dysplasia	10	Recurrent severe bronchospasm
BPD with patent ductus arteriosus	4	Pneumonia
BPD with severe reflux	5	Severe bronchopneumonia
Bronchiolitis obliterans with gastro-oesophageal reflux	12	Severe bronchopneumonia
Cardiac lesions		
Down syndrome with atrioventricular septal defect	9	Pneumonia
Tetralogy of Fallot, esophageal atresia, pulmonary artery sling, tracheomalacia	12	Severe intractable bronchospasm
Pulmonary hypertension and PDA	9	Aspiration pneumonia
Congenital abnormalities of the respiratory system		
Severe laryngomalacia with failure to thrive	5	Pneumonia
Hypoplastic left lung with tracheal ring	10	Cor pulmonale
Cerebral palsy with tracheobronchomalacia	18	Pneumonia
Congenital lobar emphysema	12	Intractable bronchospasm
Intraspinal haemorrhage	12	Severe pneumonia
Wilson Mikity syndrome	5	Severe pneumonia

underwent fundoplication. Three patients had severe bronchopneumonia, one patient had tracheo-oesophageal fistula with tracheomalacia, one patient had congenital tracheobronchomalacia, one patient had panhypopituitarism with recurrent pneumonia and one child suffered from severe Steven Johnson Syndrome. Seven of these patients were ventilated during the initial stage of their disease. At the time of writing, seven patients remained oxygen dependent ranging from two months to 1.5 year. The median duration these patients were on home oxygen therapy was 19 months (Q1:1, Q3: 48). Two patients died at the age of one from severe bronchopneumonia.

A total of 14 patients died while on this program. Table II shows the types of cases and causes of death in these patients. The majority of deaths were due to chest infection or to the natural process of the disease such as Jeunes's asphyxiating thoracic dystrophy or Down syndrome with atrioventricular septal defect. None of the deaths were related to the long-term home oxygen therapy using the oxygen concentrator.

Discussion

Home oxygen therapy may be initiated in children who developed respiratory failure but who have the potential for clinical improvement. Supplemental oxygen, allows early discharge from the hospital. The oxygen therapy may be weaned off as the underlying conditions causing the respiratory failure improved over time such as BPD and bronchiolitis obliterans.

Recent studies in infants with BPD have shown alterations of pulmonary artery pressures, heart rate variability, alterations in respiratory drive in response to oxygen and abnormal sleep patterns[7].

Harris and Sullivans[7] documented sleep disturbances such as increased numbers of arousals and decreased rapid eye movement sleep in neonates with chronic lung diseases. Although supplemental oxygen does not significantly alter oxygen saturation levels, it improves sleep architecture which is important for normal growth[8].

Home oxygen therapy is also to permit appropriate weight gain. A study conducted in 22 infants with chronic lung diseases demonstrated that the administration of low flow oxygen (0.13 - 0.25l/min) to maintain oxygen saturation of >93% was associated with normalization of weights[9]. Infants with BPD respond to even small changes in oxygenation with a significant change in pulmonary artery pressure and/or pulmonary vascular resistance. A cardiac catheterization study of infants with chronic lung showed that increasing SaO2 from 82% on air to 93% by oxygen administration reduced the pulmonary vascular resistance by 50%[10]. Oxygen supplementation in this group of children may prevent them from developing pulmonary hypertension[11].

Regardless of home oxygen therapy, these children had an increased rate of re-hospitalisation for respiratory illnesses[12]. The risks of respiratory tract infections in this group of children are dependent on the severity of the airway damage, their nutritional status and more importantly their home environment, particularly the presence of smokers and overcrowding. It is not advisable for these children to be looked after in day care centres to minimize the risks of intercurrent infections.

A second category of children who may benefit from home oxygen therapy are those who with progressive or stable diseases that are unlikely to improve. These include children with neuromuscular disorder, interstitial lung disease and pulmonary hypertension. These children are no longer able to maintain adequate oxygenation or ventilation. Oxygen therapy is used primarily to improve the child's quality of life.

A third group of children who may require home oxygen therapy are those with obstructive sleep apnoea syndrome (OSAS). In children OSAS may be associated with disorders that cause an anatomically small or abnormal upper airway such as Apert's syndrome and obesity.

Recurrent episodes of hypoxaemia and hypercarbia may affect the cardiovascular system as well as growth and behaviour. Supplemental oxygen has been shown to improve oxygenation in children with OSAS[13]. They may also need nasal continuous airway pressure support to maintain airway patency by splinting the nasopharynx during sleep.

In conclusion, home oxygen therapy is an important aspect of ambulatory care in children with chronic respiratory disorders. This is a program that needs to be expanded by all paediatricians that are involved in the ambulatory care of children with chronic respiratory insufficiency.

The current program in Institute Paediatric, Hospital Kuala Lumpur, which was first started by the UKM Paediatric Respiratory Unit, was initially sustained by donations from charitable organisations. However last year we were able to buy four concentrators from hospital funding.

References

1. Karen Z. Voter, Katren Chalanick. Home oxygen and ventilation therapies in pediatric patients. Curr Opinion Pediatri 1996; 8: 221-5.
2. Silva DT, Hagan R, PD Sly. Home oxygen management of neonatal chronic lung disease in Western Australia. Pediatr Child Health 1995; 31: 185-88.
3. Pilmer S. Prolonged mechanical ventilation in children. Pediatr Clin North Am 1994; 41: 473-12.
4. CT. Deng, BHO Azizi, M.Z. Norzila, Yeoh NM, Zulkifli HI. Home oxygen therapy in children. Experience with four cases. Annual Paediatric Congress of the Malaysian Paediatric Association June 1995.
5. Hardy KA, Schidlow DV, Zaer N. Obliterative bronchiolitis in children. Chest 1993; 3(1988): 460-6.
6. Mcnamara JJ, Urschel HC, Ardnt JH, Ulevitch H, Kingsley WB. Idiopathic unilateral hyperluscent lung, the Swyer James Syndrome. Ann Thorac Surg 1969; 7: 351-6.
7. CF Poets. When do infants need additional inspired oxygen? A review of the current literature. Pediatri Pulmonol 1998; 26(6): 424-8.
8. Harris MA, Sullivan CE. Sleep pattern and supplemental oxygen requirements with chronic neonatal lung disease. Lancet 1995; 345: 831-2.
9. Groothius JR, Rosenberg AA. Home oxygen promotes weight gain in infants with bronchopulmonary dysplasia. Am J Dis Child 1987;141: 992-5.
10. Abman SH, Wolfe RR, Accurso RJ, Koops BL, Bowman CM, Wiggins JW. Pulmonary vascular response to oxygen in infants with severe bronchopulmonary dysplasia. Pediatrics 1985; 75: 80-4.
11. Baraldi E, Carra S, Vencato F, Fillipone M, Trevisanuto D, Milanesi O, Pinello M, Zanardo V. Home oxygen therapy in infants with bronchopulmonary dysplasia: a prospective study. Eur J Pediatr 1997; 156: 878-82.
12. Chye JK, Gray PH. Rehospitalisation and growth of infants with bronchopulmonary dysplasia: a matched control study. J Paediatr Child Health 1995; 31: 105-11.
13. Marcus CL, Carrol JL, Bamford O, Pyzik P, Loughlin JM. Supplemental Oxygen during sleep in children with sleep disordered breathing. Am J. Crit Care 1995; 152: 1297-301.