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# Home mechanical ventilation in children: indications and practical aspects<sup>1</sup>

## Summary

An increase in the number of paediatric patients dependent on long-term or home mechanical ventilation has led to growing interest in this topic. Primary care physicians are becoming increasingly involved in teamwork with paediatric intensivists and pulmonologists in the care of such children. There are many causes of chronic respiratory failure and different options for provision of ventilatory support at home. Awareness of the possibilities of long-term ventilation is essential in organising discussion of these therapeutic modalities with chronically ill children and their parents prior to any respiratory compromise precluding an elective decision. Home ventilation offers the best option for these children's psychosocial development, social integration and quality of life. Many ventilator-dependent chil-

dren can attend school. The main problems of paediatric home ventilation are its negative psychosocial impact on family life, limited home care resources, the financial burden, differences in ethical perception by the local community and the fact that the equipment is usually designed for adults. Unfortunately, only scant data are available on the number of home-ventilated children in Switzerland. The creation of a patient organisation dedicated to support of families with ventilator- or technology-dependent children may be helpful in mastering some of these problems.

*Keywords:* congenital central hypoventilation syndrome; chronic respiratory failure; neuromuscular disorders; non-invasive ventilation; positive pressure mask ventilation; tracheotomy

## Zusammenfassung

Fortschritte in der neonatologischen und pädiatrischen Intensivmedizin führen zu einer Zunahme von Kindern, die von einer Langzeitbeatmung abhängig sind. Es gibt viele Indikationen zur Heimbeatmung beim Kind und verschiedene Optionen, wie diese durchgeführt werden kann. Hausärzte werden in zunehmendem Masse in die Betreuung von heimbeatmeten Kindern involviert. Ist eine chronisch-respiratorische Insuffizienz absehbar, sollte die Option «Heimbeatmung» so früh wie möglich und vor Auftreten einer akuten Dekompensation mit den Patienten und deren Eltern diskutiert werden, um den Betroffenen Zeit für einen

elektiven Entscheidungsprozess zu geben. Die Beatmung zu Hause bietet die beste Option für die psychosoziale Entwicklung, die soziale Integration und die Lebensqualität von beatmungsabhängigen Kindern. Am meisten Schwierigkeiten bei der Heimbeatmung verursachen die negativen Auswirkungen auf das Familienleben, die beschränkten Pflegekapazitäten, die finanzielle Belastung, die unterschiedliche Akzeptanz in der Gesellschaft sowie die Tatsache, dass die Geräte in der Regel für Erwachsene konzipiert wurden. Leider gibt es keine umfassenden Daten über die Population der heimbeatmeten Kinder in der Schweiz. Eine

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designierte, nationale Patientenorganisation mit dem Ziel, die Interessen dieser Familien zu vertreten, wäre hilfreich, um einige der angesprochenen Schwierigkeiten zu meistern.

*Keywords: Tracheotomie; Maskenbeatmung; nicht-invasive Beatmung; kongenitales zentrales Hypoventilations-Syndrom; Zwerchfell-Pacing*

## Introduction

Advances in neonatal and paediatric intensive care together with improvements in early diagnosis and intervention of respiratory failure have resulted in a higher survival rate, but have also resulted in a new treatment population, a population of children dependent on long-term mechanical ventilation. Increasing importance is being placed to transfer these children from paediatric intensive care units to the home environment or at least to less acute areas [1, 2]. Improvements in sophistication and miniaturisation of ventilatory and monitoring equipment allow to accommodate home care needs and mobility of technology-dependent patients. It has become evident that providing ventilatory support at home offers the best option for the child's psychosocial development and quality of life [3]. Finally, the direct cost of home care is usually lower than that of hospital care. Nevertheless, according to a recent survey in the United Kingdom a significant number of children requiring long-term venti-

lation remain hospital bound, although major advances in ventilatory and monitoring equipment allow even small infants and children to be ventilated safely at home [4]. The major obstacles which delay hospital discharge are failure to recruit qualified nursing staff to care for the child at home or unsuitable social conditions [5].

Most experience with managing children on home ventilation is relatively recent and usually confined to a few centres. There is, however, a growing interest in this topic, since primary care physicians are becoming increasingly involved in the care of such patients. It is not the intent of this report to provide a comprehensive review, but to discuss some of the issues and problems involved with ventilating children at home. Unfortunately, there are hardly any data available on the number of infants and children on home ventilation in Switzerland.

## Indications for long-term ventilation in paediatric patients

Chronic respiratory insufficiency can be defined as a life-affecting or growth-affecting sit-

**Table 1**

Causes of chronic respiratory insufficiency in paediatric patients.

increased respiratory load
chronic cardiopulmonary disorders
upper airway obstruction: vocal cord paralysis, craniofacial syndromes
chronic lung disease: bronchopulmonary dysplasia, lung hypoplasia, cystic fibrosis
congenital heart disease
skeletal deformities
kyphoscoliosis, thoracic wall deformities
ventilatory muscle weakness
neuromuscular disorders
spinal muscular atrophy, muscular dystrophies (e.g. Duchenne), others
polio
phrenic nerve paralysis
spinal cord injury (above C-3)
failure of control of ventilation
congenital central hypoventilation syndrome (Ondine's course)
acquired: trauma, tumour, surgery, haemorrhage, radiation
myelomeningocele: Arnold Chiari type II

uation due to a long-term problem with oxygenation and/or ventilation. Respiratory dysfunctions supported or treated with long-term ventilation in paediatric patients can be divided into three pathophysiologic categories: excessive respiratory load, ventilatory muscle weakness and failure of central ventilatory control (table 1).

Pulmonary diseases that lead to chronic respiratory insufficiency in infancy, such as bronchopulmonary dysplasia, may improve with age and allow withdrawal from mechanical ventilation. It is important to realise that the respiratory system of infants is immature and prone to respiratory failure, but has a great rehabilitative potential as a result of the growth in small airway size and the increase in alveolar number during the first decade of life [6]. Long-term ventilation allows these children to grow, preserves physiologic function and prevents further damage from respiratory deteriorations. In older paediatric patients with chronic lung disease, such as cystic fibrosis, home ventilation may be used to improve sleep-related hypoxaemia and hypercapnia

and as a short-term bridge to transplantation [7–9].

Similar expectations as for the growth and developmental potential of the lung parenchyma can be made for the infant chest wall which will become stiffer and more stable with increasing age. Personal experience relates to a boy with Jeune asphyxiating thoracic dystrophy who was successfully decannulated in later childhood. The most common disorders for which children are ventilated at home are neuromuscular disorders and the congenital central hypoventilation syndrome (CCHS). In children with neuromuscular disorders, nocturnal non-invasive positive pressure ventilation may prevent respiratory deteriorations secondary to atelectasis or respiratory infections and decrease hospitalisation rates. Ethical concerns regarding the institution of long-term invasive ventilation through a tracheostomy in infants and small children with progressive neuromuscular disorders relate to the fear of prolonging suffering from a miserable and unfavourable disease. Intermittent non-invasive positive pressure ventilation may offer a valuable alternative and serve as a comfort measure to relieve anxiety from hypoventilation and hypoxaemia

with probably little impact on long-term survival of such children. Death is usually a result from insufficient coughing and clearing of secretions rather than from insufficient ventilation in patients with progressive respiratory muscle weakness.

The congenital central hypoventilation syndrome (CCHS) is a rare disorder characterised by the absence of adequate autonomic control of respiration, but is one of the more common indications for paediatric home ventilation. Most of these infants need ventilatory support since birth and demonstrate adequate ventilation during wakefulness but not during sleep. Nevertheless, CCHS presents in variable degrees of severity ranging from complete apnoea during sleep and severe hypoventilation during wakefulness to mild hypoventilation during quiet sleep. The most severely affected infants require 24-hour ventilatory support since birth. Some may acquire awake hypoventilation at the age of 2–4 years as a result of an increase in physical activity. Today children with CCHS have a reasonable long-term medical and psychosocial prognosis and will usually be offered the necessary therapeutical options without hesitation [10].

## Initiation and management of long-term ventilation

Usually the decision to initiate long-term ventilation is made non-electively in the neonatal period or in later life after presentation with acute respiratory failure. In many circumstances there is no opportunity to discuss the possibility of home ventilation in advance and there is usually little choice for the child and its family.

Unfortunately, even when the need of ventilatory support could be anticipated early as in older paediatric patients with progressive disorders, long-term ventilation and end-of-life care is often not discussed with the patient and the family prior to the first catastrophe. Patients at high risk for respiratory failure (especially older children with neuromuscular disorders, musculoskeletal disease or cystic fibrosis) and their parents should have the opportunity to thoroughly review and discuss the

potential need for ventilatory assistance prior to its initiation. This may prevent inappropriate avoidance or involuntary installation of long-term mechanical ventilation due to misconception or inadequate expectations. Physicians taking care of chronically ill children should provide sufficient information on the benefits, risks and burden of long-term mechanical ventilation to enable the patient and the family to participate actively in the decision-making process prior to any respiratory disaster.

There are many practical issues relating to the management of children requiring chronic ventilatory support (table 2). The relative infrequency with which this is instituted means that it is best done by centralised and experienced institutions. The decision to send a ventilator-assisted child home must be made in cooperation with its family. Strategies prior to discharge must address the issues of providing equipment and supporting personnel, family education, arrangement of the home environment, rehabilitation services and follow-up examinations (fig. 1).

The cornerstone of a successful discharge of the ventilator-dependent child is medical, psycho-

**Table 2**

Main problems involved in home ventilation of children.

equipment usually designed for adults

psychosocial impact on family life

limited home care resources

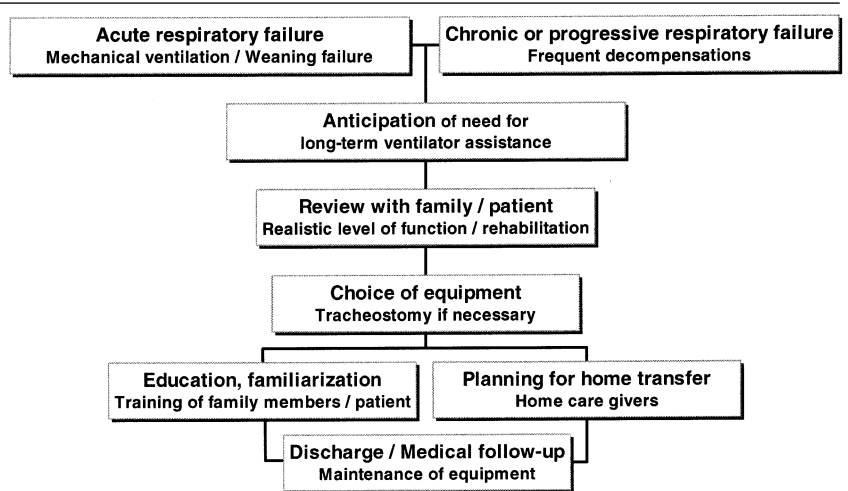
financial burden

perception by community

ethics

**Figure 1**

Outline of management for long-term mechanical ventilation.



logical and psychosocial stability. The child is never too young to go home, but the arrangement of a functional environment requires time and patience, especially when discharging children to remote areas with limited home care resources. Ventilator requirements should be stable and major diagnostic evaluations or therapeutic interventions should be completed. Treatment of the underlying disease should be maximised in terms of pharmacological and nutritional support. The main difficulty, however, is not so much clinical as social. The key to successful home ventilation usually lies in the ability to provide adequate social support for continuation of a functional family life at home. The level of support at home will vary with each child and family and depends on many different contributing factors such as the age of the child, the level and mode of ventilatory assistance, the presence of physical handicaps, the ability of the family to cope, the number of children in the household, work commitments and other social and environmental issues. The level of support is best determined together with the parents and should be revised on a regular basis to account for changes in disease severity or family structure. A tight collaboration of physicians, nursing staff, social workers and home care providers is required to prevent unnecessary delays in the discharge process. Unfortunately, in some children discharge is delayed due to failure to recruit sufficient nursing staff [5]. Discharge may occa-

sionally be impossible due to family unwillingness, inability to cope or other social problems. Family members and home care providers are best trained in the hospital prior to discharge using the patient's own home equipment. All persons need to be adequately prepared for providing routine and emergency care of the child outside the hospital. Parents and caregivers of tracheotomised children should be trained to change tracheostomy tubes and recognise and correct the most common problems such as tube dislocation or tube obstruction. Training also includes competence in basic cardiopulmonary resuscitation and the correct use of supplies and equipment. Parents and professional home care providers are best trained separately. Parents should not be responsible for the initial training of home care nurses. They will, however, be involved in the training process at home. Equipment companies or specialised community services can provide equipment maintenance and help in case of malfunction. In certain circumstances, gradual discharge or a trial run at home may be useful to decrease initial fears and uncertainties and to document unexpected problems. Gradual discharge may consist of a day at home, followed by one or more trial weekends before final discharge. We recommend to build a core team to communicate with the family and to discuss problems and changing needs of the patient with the respective services.

## Methods of home or long-term ventilatory support

Different ventilation techniques are available for long-term or home ventilatory support in children. The decision which technique to choose depends upon the underlying disease, the age of the child, the duration for which ven-

tilatory assistance is required, the acceptability to the patient, local experience, availability of equipment and cost. The choice of equipment should also be based on the principle of using the simplest technology capable to support the

patient for life-style needs. The following methods will be briefly discussed:

- a positive pressure ventilation via tracheostomy,
- b non-invasive positive pressure mask ventilation,
- c diaphragmatic pacing,
- d negative pressure ventilation.

#### **Positive pressure ventilation via tracheostomy**

Positive pressure ventilation via tracheostomy is the traditional method of home and long-term ventilation in infants and small children. A tracheostomy provides the safest access to the airway when ventilating children at home. Tracheostomy tube sizes are usually chosen as small as possible to ensure adequate ventilation and to allow a leak for the maintenance or development of speech. This may make it impossible for volume-targeted ventilators to maintain a constant minute ventilation, because the tracheostomy leak is usually relatively large and variable. The newer generation of pressure-control ventilators handle the leak problem much better. Traditional BiPAP ventilators designed to treat obstructive sleep apnoea in adults are neither suitable nor designed to provide full ventilatory support in tracheostomised children.

Although an increasing variety of portable ventilators suitable for home ventilation is available on the market, basically all home ventilators are not designed to operate below a tidal volume of 50–100 ml. This limitation not only poses a problem to the older generation of volume-targeted ventilators, but also to the newer pressure-control ventilators when ventilating children below 10–15 kg body weight. Since tidal volume measurements are part of the ventilator alarm system, this may be constantly triggered by tidal volumes below the critical range. The insertion of a pressure limiting pop-off valve into the inspiratory circuit of the ventilator provides an artificial and constant leak and helps to overcome the alarm problem in pressure-control ventilators. This measure also changes the volume-targeted ventilator into a time-cycled, pressure-limited ventilator and facilitates its use in small children. Tracheostomy is associated with a significant number of disadvantages and complications. There is a need for permanent and close surveillance to prevent cannula-related deaths from tracheostomy tube displacement or obstruction [11]. Frequency and severity of lower

respiratory tract infections are increased due to the loss of the protective role of the upper airway. Language acquisition and articulation is impaired. Swallowing dysfunction and disturbances in olfaction are also common problems in tracheostomised children [12]. An effective home care programme requires parental training in tracheostomy care, the use of ancillary equipment and infant cardiopulmonary resuscitation [13].

#### **Non-invasive positive pressure mask ventilation**

Non-invasive positive pressure mask ventilation is an elegant technique to provide ventilatory assistance without the need for a tracheotomy. It has become an important tool in adult pulmonary medicine for temporary or nocturnal ventilatory assistance. At present, there is little experience with long-term nasal mask ventilation in infants and small children, but there is an increasing number of reports describing its successful use in paediatric patients [14, 15]. Non-invasive positive pressure mask ventilation may be considered in children if ventilatory requirements do not exceed 12–14 hours per day and is best attempted on a nocturnal basis. The best candidates for long-term nasal ventilation are older children with chronic respiratory failure due to stable or slowly progressive neuromuscular disorders, chronic lung diseases, chest wall deformities or hypoventilation syndromes.

At present there are hardly any published reports on the use of nasal mask ventilation in infants and young children at home. More clinical research has to be performed before this technique can widely be recommended in this age group. There are also several disadvantages or dangers associated with using a mask to support ventilator-dependent infants and small children at home. Ventilation may be inadequate because of leakage through the mouth or the mask. Upper respiratory tract infections are common during the first few years of life. The lack of a secure airway may result in anxiety, more frequent hospitalisation or even intubations during respiratory deterioration caused by otherwise simple respiratory tract infections. Another serious concern with using a mask to ventilate infants and children at home is that mask pressure will affect facial growth and result in mid-facial hypoplasia [16]. Therefore, we personally favour to ventilate infants and young children primarily by tracheostomy and switch them to mask ventilation in later

childhood. Other problems of mask ventilation include pressure sores and air swallowing which are usually easy to deal with.

Arguments in favour of mask ventilation is the avoidance of tracheostomy and all its disadvantages and complications, such as recurrent tracheitis/bronchitis, laryngotracheal damage, tube obstruction or displacement, speech impairment and cosmetic handicap. It may be expected that using non-invasive forms of ventilation may improve quality of life, facilitate social integration and school attendance, and lower cost when compared to ventilation via tracheostomy. In certain circumstances, non-invasive mask ventilation may also be the best of all options for infants and young children. A fiberoptic evaluation of the upper and lower airways should always be performed before starting or switching children to long-term mask ventilation to assure upper airway patency. It is this author's experience that adenotonsillar hypertrophy is a common feature of children with long-term tracheostomies and needs to be assessed prior to decannulation and/or switching to non-invasive ventilation. Adenotonsillectomy may occasionally be required [17].

### Diaphragm pacing

Diaphragm pacing is based on electrical stimulation of the phrenic nerves to generate breathing by activation of the diaphragm [18, 19]. It is commonly performed bilaterally and has become an important alternative mode of respiratory assistance for children who require respiratory support during both wakefulness and sleep [20]. The patients wear an external radiofrequency transmitter over an implanted receiver. A stimulating current is induced without the need for any transcutaneous wires. Most experience with diaphragmatic pacing comes from children with CCHS or adult patients with high cervical cord injury. In children with the most severe form of CCHS pacing is commonly used at day and positive pressure ventilation through the tracheostomy at night. There are concerns that 24-hour pacing may induce fatigue or permanent injury to the diaphragm or phrenic nerves in children. Diaphragmatic pacing is contraindicated in patients with abnormal respiratory mechanics, primary myopathies or phrenic nerve lesions.

Most children require a tracheostomy or continuous positive airway pressure to prevent upper airway obstruction during electrically stimulated inspirations. The underlying mechanism relates to the absence of normal inspiratory activation of the upper airway abductor that normally precedes activation of phrenic nerves and serves to dilate and stabilise the upper airway [21]. Nevertheless, decannulation and successful discontinuation of tracheostomy tubes have been reported in selected patients, usually older children or adult patients. The advantage of phrenic nerve pacing is that the transmitter that provides electrical stimulation is small and easily portable, thus allowing more mobility. The disadvantages are that it is expensive, requires surgical implantation and lacks an alarm system in case of malfunction. Managing patients with diaphragmatic pacers requires expertise with implantation, gradual conditioning of the diaphragm and adjustment of the electrophrenic pacer to provide adequate ventilation.

### Negative pressure ventilation

Negative pressure ventilation is one of the oldest forms of ventilatory assistance and was widely used during the polio epidemic. Many types of negative pressure devices are available where either the whole body from the neck downwards or just the trunk is enclosed in a rigid container (iron lung, poncho, cuirass etc.). These devices provide adequate ventilatory assistance particularly for patients with chronic respiratory failure due to slowly progressive neuromuscular disorders or alveolar hypoventilation syndromes. Most published experience with negative pressure ventilation in paediatric patients comes from the United Kingdom [22]. Despite the recent progress in non-invasive ventilation techniques, modern negative pressure machines may give the respiratory physician and his patients a welcome alternative. Inefficiency has been reported in patients with distorted chest walls and upper airway obstructions [23]. A major disadvantage is that medical and nursing staff are relatively unfamiliar with this therapeutic modality. In addition, there is a tendency of the upper airway to collapse causing upper airway obstructions by a similar mechanism as in diaphragmatic pacing.

## Additional equipment for paediatric home ventilation

The equipment for successful paediatric home ventilation has to ensure safety and mobility of the patient in his specific social environment. For most patients a pulse oximeter is sufficient for adequate home monitoring, although capnography may be useful in certain circumstances. We favour to supply the patient with a small portable and more sophisticated stationary pulse oximeter. Although ventilator equipment failure is rare and hardly a serious problem, a second back-up ventilator is required for safety reasons in almost all ventilator-dependent children [24, 25]. We favour to equip wheelchairs or strollers with a secondary ventilator to facilitate mobility of the patient and the family. Both ventilators should be used alternatively to assure that both remain func-

tional. A humidification system should at least be incorporated into the stationary ventilator used at night. Further equipment includes a portable suction with suitable catheters in the case of tracheostomy, an autoclave to clean the ventilator tubings and a self-inflating resuscitation bag with mask. It is evident that spare interface equipment such as masks or tracheostomy tubes should be readily available. If additional oxygen is required in ventilator-dependent children, we favour the installation of liquid oxygen tanks at home. Small portable cylinders allow for mobility of the oxygen-dependent child. Further equipment may be required depending on special individual needs and social circumstances (fig. 2 and 3).

### Figures 2 and 3

Providing mobility to children on long-term ventilation often requires imagination for individual solutions.



## Regular follow-up of children on home mechanical ventilation

Short-term hospitalisations are usually required for regular follow-up examinations of children on home mechanical ventilation to adjust the ventilatory support to the changing anatomy and physiology of the respiratory system during growth. Such hospitalisations also facilitate the coordination and collaboration of all other disciplines involved in the care of these families. These controls not only address the evaluation of the respiratory system, but also include regular nutritional, developmental, neurologic and psychosocial assessments. Depending on the child's underlying disease other specialities may need to be involved for a comprehensive multidisciplinary approach in the care of these children.

We favour to schedule the first follow-up examination not later than 3 months after discharge, for which the child is usually hospitalised for 1–2 days. The subsequent frequency depends on the individual needs of the family and the child. We recommend to admit home-ventilated children every 6–12 months for overnight monitoring and a multidisciplinary check-up. Older children with chronic disease and stable ventilatory requirements are followed on an annual basis. Other regular assessments include ECG, echocardiography, pulmonary function testing and fiberoptic evaluation of the upper and lower airways of all tracheotomised children, usually performed every one or two years. Patients with rapidly chang-

ing conditions may require more frequent controls.

Occasionally families have to be relieved from the burden to take care of these children to enable vacations and other activities. Physicians involved in the care of home-ventilated children should be able to offer parents and caregivers such "holiday hospitalisations" to ease the psychosocial burden on their family life and partnerships, if necessary. All efforts to provide

such "holiday care" of stable home-ventilated children in local, nearby hospitals should be greatly encouraged. Providing such care in facilities without the security of an intensive care unit remains a major problem in our country. Efforts to keep ventilated children at home are only successful if one takes meticulous care to keep the family environment healthy and functional.

## "Pro and Cons" of paediatric home ventilation

The benefit of home ventilation includes a better quality of life, optimising rehabilitative potential and participation in society such as attending school. Placement of home-ventilated children in our school-system is not very common, but has successfully been instituted in selected patients. Whenever possible a ventilator-dependent child should be educated in as normal a school setting as possible. There is evidence that installation of ventilatory support on a selective basis leads to lower hospital re-admission rates in patients with neuromuscular disorders [26].

Today's technical ability in critical care to extend life using mechanical ventilation often leads to the question of whether it should be done rather than the question of whether it can be done. Different ethical perceptions in the community may cause ongoing discussions and problems of acceptance for families and care providers of home-ventilated children. Besides ethical concerns, there is insufficient published experience on long-term ventilatory support of patients under two years of age, except for those with CCHS. Nevertheless, providing non-invasive ventilatory support at night or

during respiratory infections can also be regarded as a comfort measure and even be justified in children with progressive disease.

The major problem of chronic home ventilation is the large psychosocial impact on the family causing loss of privacy and limited social lives. Improved resources of community health teams could help to facilitate the complicated discharge process. It is the opinion of this author that home care providers do not necessarily need a nursing qualification as long as they are adequately trained by nursing staff and physicians. Such measures may become necessary to overcome shortcomings in nursing resources and may also decrease home-care costs. Sometimes the care of handicapped and ventilated children becomes more expensive at home than institutional alternatives. Nevertheless, it is not appropriate for any ventilator-dependent child to grow up in a hospital environment. Finally, a patient organisation dedicated to support of families with ventilator- or technology-dependent children may be helpful in mastering some of the financial and social problems these families have to deal with in our country.

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