〔シンポジウム：小児呼吸管理の最近の進歩〕

Care of ventilator-dependent children

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在宅人工呼吸患者の管理

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以下の要約は昭和61年7月に行われた第8回人工呼吸研究会でのロバート・ケトリック先生の口演記録を基に作成した内容です。講演内容の全文は大阪大学西村匡司氏の御努力で作成されました。スライドの大半は情況描写的な内容であり，紙面の都合上割愛させて頂きました。

要　約

集中治療の発達に伴い，急性期に人工呼吸管理を受けた症例の人工呼吸管理が長期化し ICU ベッドの大半を占める情況が各地の ICU で見られつつある。こうした人工呼吸依存の患者を在宅で管理しようとする試みは，医療行政の経済性の面からも，患者個人および家族の情緒の面からも意義のあることであり，米国連邦政府もクープ医務長官の下に在宅管理検討委員会を設け政府レベルの対応を検討することになり，現在その準備をすすめている。

小児で在宅人工呼吸の対象となる患者には二つのグループがある。すなわち，慢性神経筋肉疾患や脊髄損傷患者などと，所謂テクノロジー依存患者と呼ばれる集中治療後の慢性呼吸不全患者群である。フィラデルフィア小児病院での経験は専ら後者が主であり，今回はこの患者群の取り扱いについて述べる。

フィラデルフィアの小児病院では1967年～1984年の間に101名の小児慢性呼吸不全患者を取り扱った。このうちの20名は死亡し81名は生存した。本年度現在の生存者は71名であり全体としての死亡率は30%である。この中で全体の54%が結果的に人工呼吸器から離脱できたことは重要な点である。死亡例を検討してみると，その大半は生後一年半未満であり，それ以上人工呼吸を続けた症例の生存期間は長いといえる。

フィラデルフィアの小児病院での退院患者の大半は4歳以下である。気管支肺異形成症を例にとると，病院での人工呼吸日数は92～1,400日平均683日，在宅人工呼吸で離脱が可能であった症例は132～1,032日で平均365日であった。

経済面ではフィラデルフィアの小児病院での ICU 滞在費年間30万～35万ドルを2万～8万と80%以上の医療費節減に役立つことが分っている。実際の患者の在宅医療への移行については講演の中で触れる（文責　宮坂勝之）。

First let me extend my gratitude and appreciation to Professor Yoshiya and Dr. Miyasaka for inviting me. Also let me extend greetings from the faculty of Children's Hospital of Philadelphia and University of Pennsylvania.

I am going to present information on a home care program for ventilator-assisted children that was developed at the Children's Hospital of Philadelphia. This first series of slides

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represents a case presentation of the kind of patient that I am talking about. This little baby was born with a complete tracheal web, a posterior laryngoesophageal cleft, imperforated anus, tracheoesophageal fistula, and ventricular septal defect. The child was born at the Pennsylvania Hospital and within two hours he was transported to the Children's Hospital of Philadelphia, where an emergency tracheostomy was done.

Over a period of five years in our hospital, the child grew and developed. Four of those five years, the child was dependent on mechanical ventilation. Much of that was related to tracheomalacia, because of an incompetent larynx after the laryngoplasty was completed. The child had recurrent aspiration, tracheomalacia from liquidification necrosis of the tracheal cartilage, and ventilator-dependency for that reason.

The child's mother was in jail because of drug abuse and a history of violence. So we became the parents for the child. While in the jail, the mother had two more children and this is one of the siblings who came to visit.

Just to give you a feeling for the kind of environment that we've tried to create in the hospital for the children who are ventilator-dependent, we make an effort to provide them with social development and to provide them with freedom from painful or unpleasant experiences.

The child grew older, became less dependent on the ventilator, but still remained dependent on the CPAP system. So a portable CPAP system was developed for the child. Do you see the child's swivel connector which is located at the tracheostomy and connected to a portable CPAP system ?

You can see the reservoir for the CPAP hanging from his back with a very long oxygen hose, sometimes 75 to a hundred feet (30 m) of oxygen hose, going back into a room around the corner in order to allow the child to become very mobile.

The child was eventually weaned off mechanical ventilatory support, but he still had no family to take him home. We were able to find a foster family for him. He was eventually discharged to the care of foster parents, an older couple whose children had grown up and moved away. You can see him using his electrolarynx. He was also very good at sign language and knew about a hundred and fifty different words with sign language. All of these are due to the program that we have in Philadelphia for ventilator-dependent children who have communication handicaps that go with being on ventilator and having a tracheostomy. The child was home for a year. His tracheostomy cannula became plugged in the presence of his foster mother. Even though she had the child home for a year and had changed the tracheostomy at least fifty times, she panicked and forgot to change the tracheostomy. By the time the rescue squad came, the child had had a cardiac arrest, and he was transferred back to our hospital with severe hypoxic encephalopathy. He died some months later.

Between 1967 and 1984, we have taken care of over a hundred one (101) children with chronic respiratory failure of infancy. We have had many more older children with chronic respiratory failure, but I am just going to talk today about chronic respiratory failure evolving from the period of infancy. Of those, we had twenty children (roughly twenty percent) who were never weaned and died, and about eighty-one percent who lived. Of the latter, seventy-one are currently living and an additional ten have died. Patients were all referred to us from outside of the newborn intensive care unit. Thirty percent of these

children died. Perhaps more importantly, fifty-four percent of these children were eventually weaned from mechanical ventilation. Overall, there is a good prognosis for this patient population for eventual weaning from the ventilator.

But also, a substantial number of those patients (thirty percent) have died. When do they die? These patients were referred to us because of the inability of primary care takers to wean them from mechanical ventilatory support. When we look at death as a function of time, most of these children who die are dying within the first 12 months of life. We can see the incidence of death rises very rapidly but then begins to plateau off. Those children who have lived beyond the first year to a year and a half are probably going to live for a substantial period of time.

We looked at our early experience between 1967 and 1976, a nine year period when we had 23 patients. Between 1967 and 1976, those patients who came to us with chronic respiratory failure of infancy had a tendency of an incidence of early death which then plateaud off. After about thirty six months, the death rate was very very low. In a subsequent two-year period, we had 27 patients. I do not know why there is a difference between these two groups. But the death slope in the later period is very steep. In addition, it also plateaus out, indicating that the children who live to three years are probably going to live for an extended period of time. Interestingly, between 1980 and 1982, we had 29 patients, and we saw a change in the death slope. Perhaps we are learning to identify these patients earlier, and to take better care of them. But there are clear differences in the rate of survival, and it looks like these patients who come to us with chronic respiratory failure of infancy are not going to die quickly. In fact they live for long periods of time. Those with muscular disease, for example, traumatic quadriplegia, may, in fact, live decades.

We have been seeing more and more patients with chronic respiratory failure of infancy. We had a flurry of them actually. These children were at the Children's Hospital of Philadelphia before I came. Earlier on, we began to see that the incidence of children admitted to our special care unit rose rather dramatically, and then admission plateaued off between 1980 to 1983. The reason for this plateau was very simple: we stopped taking patients. We have a total of 33 beds in our pediatric intensive care unit, and by this time more than half of the beds in the pediatric intensive care unit were occupied by infants with chronic respiratory failure. We had very active surgery programs (neurosurgery, cardiac surgery, craniofacial surgery and so forth), and we were not able to do the elective surgery that was a part of the hospital's mission. Further, we were transferring acute care patients away from our emergency room to other emergency rooms, because we didn't have beds available to admit these children to the intensive care unit. So we stopped taking chronic patients. With the development of a home care program for chronic respiratory failure in infancy, we began to move more patients through the system. Then we began to accept more admissions for chronic respiratory failure.

This slide simply shows the yearly discharge rate for ventilator-dependent children at the Children's Hospital. In 1981 we were back down to the lowest level; in each successive year, we have an increased number of discharged patients to the home care program.

Let me try to calculate the patients population for you. This is a summary of three recent articles; one by the group at the Texas Institute for Research and Rehabilitation one by

Allen Goldberg's group at the Children's Memorial Hospital in Chicago, and one by the group at the Massachusetts General Hospital. All of these recent reports are about children who have been transferred to home care programs. What is interesting about these reported group is that there are no newborns who have been transferred home ; 75 % of the children were more than 4 years of age at the time they were transferred to home. Most of these children had neuromuscular disease, general myopathy, Werdnig-Hoffman syndrome, or traumatic quadriplegia. So we are looking pretty much entirely at a patient population with neuromuscular disease and traumatic spinal cord injury with only a few cases of pulmonary disease.

This is in contrast to the experience at Children's Hospital of Philadelphia. More recently we have seen a different pattern of discharges. Most of our patients are newborns and older infants with bronchopulmonary dysplasia. About 75% of the children that we have discharged have been under 4 years of age. So the pattern that seems to be developing in the United States is that once we recognize chronic respiratory failure of infancy regardless of the cause, we are beginning to discharge at a much earlier age where as previously children were discharged after 4 years of age, The more recent experience is that we are going to discharge children in the first one to two years of life. The earliest discharges for chronic respiratory failure of infancy have been for children seven and six months of age.

We have had a significant number of children with neuromuscular disease, general myopathy, myotonic myopathy (including sheath myopathies,) a small number of spinal cord injuries, some central hypoventilation syndromes, and a few children with severe encephalopathies either hypoglycemic or hypoxic origin. Because these children with neuromuscular disease all have good central nervous system prognosis and cognitive potential, these chilbren can grow up to be well educated and bright children. The biggest group we have seen now is the group of bronchopulmonary dysplasia, a disease that may be preventable. Also, we had another group of children with tracheomalacia. Tracheomalacia is also an acquired disease perhaps because many of the patients with bronchopulmonary dysplasia have recurrent tracheitis with damage to the cartilage skeleton. Particularly in those children who have laryngeal dysfunction and chronic alkaline aspiration, a high incidence of tracheomalacia is observed. So there is a spectrum of diagnoses that produces chronic respiratory failure of infancy. The group that seems to be growing rapidly in the United States is the group with bronchopulmonary dysplasia.

This is a review or our experience with a home care program for patients with bronchopul monary dysplasia. We have now discharged sixteen of these children to the home environment. Thirteen of the children have been weaned off mechanical ventilation and eleven children have been decannulated. Three of our original patient population still require mechanical ventilation. Our initial approach to bronchopulmonary dysplasia was not to send them home. We initially started only sending children home with severe encephalopathies and others who we thought we could never get off the ventilator. We sent them home because the parents wanted them home and we thought they could provide good care at home. We did not originaly send children home with bronchopulmonary dysplasia because of their age, but also because we thought that this was a self-limited disease

from which all of these children would recover with proper hospital environment. The hospital care program for this particular group seemed to be successful, even though the children were admitted with cor pulmonale, right ventricular hypertrophy, dilatation and strain. The hospital based program was successful in reversing those phenomenae. So we thought that since there was light at the end of the tunnel, and the children could be weaned, we would keep them in the hospital and keep them in the optimal medical environment. We subsequently found out the hospital is not the optimal medical environment for children with bronchopulmonary dysplasia and chronic ventilator dependency. Our current experience would suggest that these children have fewer infections, their pattern of nutritional intake and growth is better, and that their socialization and developmental progress is better at home. Once the parents express an interest in taking their child home, it is our practice to move these children to home care as quickly as possible. Thws by around six months of age, we are looking to a home care program.

For the bronchopulmonary dysplasia patients, the duration of mechanical ventilation in the hospital averages 683 days, with a range between 92 days up to 1,400 days. The duration of mechanical ventilation at home for those children who have been weaned is about 365 days with the range being 132 to 1,032 days.

What is happening in the United States now is that the funding by the third party payment system is driving patients from the hospital to home care. The question of when you should medically send these children home still exists. We bring the child into the intermediate care system to offer program of developmental nutritional support while ventilatory support is optimized for intermediate care. These children are transferred to the unit because it has not been possible to wean them off from the ventilator. We do take it upon ourselves to focus on weaning. We take it upon ourselves to make sure they are well oxygenated. We make sure they are free of metabolic acidosis and then move on to look at nutrition and developmental care. Once the parents are sufficiently comfortable with taking care of their child, and once they approach us about taking their child home, then we look at criteria for sending these children home. We try to make the choice as much as possible a family decision. This is something the family wants to do. Prior to the union, we have done a whole number of things to reinforce the family's confidence in taking care of their own children. I want to review our own criteria. First, of all, we insist that they are medically stable. I do not particularly care what Fio_2 is or what the degree of mechanical ventilatory support is. What we try to establish is whatever level of support they require for stability so that they do not have wide swings of saturation or desaturation from a baseline or wide swings of end tidal CO_2. We want to make sure that they are gaining weight at a regular rate and that they are free of infection.

The second criteria is that the parents are well-informed about what the implications of home care are to their finances and to their life style.

We make sure that the support system is practical. Some children live in cities, some children live out on farms, in rural isolated areas. In all cases, we make sure that there is a support system in that community to take care of the child on the ventilator. We will not send the child into a system where

support services are not available.

The fourth criteria is the family's willingness to take care of the child. The family must go through a hospital based program of education that prepares them to effectively learn the care of the child. Once they have completed that program and passed early essential markers, then we define them as willing to take their child home.

The last factor is money. We have to have sufficient money to purchase the equipment, of monitoring, and nursing services at home. For most of the children with bronchopulmonary dysplasia, we insist that there be 16 hours of nursing care in the home to start with. Once the family settles in and becomes more comfortable with the child, then if they choose to increase the amount of nursing, if their child's care is more complicated, then we will look to provide 24 hours of nursing care at home. If the family wants to adjust downwards, then we adjust downwards. So we leave the level of nursing care to be provided at home as a function of the child's needs. That is peculiar to our program in Pennsylvania. The rules standards vary tremendously from state to state. There is not yet a federal program or federal standards. We are working to make that available. So my remarks really apply to the services that are just being extended from the Children's Hospital of Philadelphia in the Commonwealth of Pennsylvania.

Now I would emphasize one other thing. Our program is clearly to transfer responsibility from the hospital staff to the family. There are a few other states that have developed programs involving case manager who is usually a nurse a social worker with a masters degree, who is substitute as a case manager dislapcing the parents to co-ordinate responsibility for care at home. Our belief is that the family is the stable source for care of their own child and all we need is the time and patience to provide an education for the family. If you do that, you can provide as much with less cost as it does when you use a manager. In regard to home care funding, we have to make the case to the third party payment system that it's cost effective. As I pointed out in the abstract, the average hospital cost for a child in our intensive care or intermediate unit varies between 300,000 and 350,000 dollars a year. If we can cut the cost by 80%, then most third party payers will consider that would be cost effective. The fact of the matter is that the home care programs we have developed are 25 to 30% of that of hospital cost. So in most cases we can cut the cash flow from the insurance company or public funding from 300,000 to 350,000 dollars down to anywhere between 20,000 to 80,000 dollars. So home care can be tremendously cost effective. We will not do it unless there is clear benefit to the child and the family. The child's safety and the family's firm committment should be confirmed. We will not do it unless we are sure that care in the home can be safe.

These children were transferred to us from other intensive care units. Many of those intensive care units are very paternalistic; the doctors and the nurses say, "we are responsible for the child, we will take care of the child, and we will come and visit the child, but the child is so desperately ill that only we can properly supervise the care." So many of the families come to us with apprehension and with fear of approaching their own child. Our goal is to step away from paternalism and move responsibility towards the family. We have a very organized fashion of "desensitizing" the family to their own child with behavior modification. We serve as role models and get them started first letting them feel

comfortable with just touching and holding their child, and providing comfort to their child. Giving their expressions of love and feeling comfortable by their child's response makes them believe that their child is appreciating the care that they give them.

Once they feel comfortable coming to the intensive care unit going to the bedside frequently, putting the bedside frame down to gain access to their child, and independently comforting their child, then we move on to the next stage. We encourage them to provide clothes and we let them get involved in activities of daily living in the hospital. They take the dirty clothes home and bring the fresh clothes in. We let them get involved in well-baby care; once they become comfortable in these areas, we gradually let them get involved in the special care needs of the child. For example while observing a nurse suctioning the child, they may hold the long suctioning catheter. We eventually move them up to suctioning the child and changing the tracheostomy by themselves.

During all of these processes, we never talk to them about taking the child home. We do not want that to be a threat to them. Our major goal is to get them to incorporate the child into the family. Once they have a sense that they can take care of their own child, then they generally come to us and say, "I understand you have money for nursing for home care. I feel comfortable with looking after my child's care. Can I take my child home?"

The third step is letting them identify the child as a part of their family. They can bring their own quilts, their own photos, and their own toys. We like it when they say, "I can take care of my child better than the new intern," or, "I can take care of my child better than the nurse on the three-to-eleven shift," or, "I do not like this being done to my child." Although some of the house officers and nurses don't like to deal with such behavior, we encourage the families to speak up and to demand change for their child. That's a strong indication that they are incorporating the child into their sphere of sphere of responsibility.

Once they have incorporated the child into their sphere of responsibility, they are much more able to accept responsibility for home care. We want them to adopt a sense of superiority about their ability to take care of their own child. We encourage them to start talking about the education and schooling of their child.

Usually after they are above and through that stage, they come to us and say they have also seen other families coming back to visit with their ventilator-dependent child and seen other children transferred to home. So it becomes much easier to develop a flow of patients in and out of the unit. After they indicate that they have an interest in taking the child home, then we put them for the first time into formal programs of education where they have to study tracheostomy care. There is written material for them, and they have to take an examination on practical aspects. They do a tracheostomy change under the supervision of a nurse on three separate occasions. The tracheostomy management skills are broken into two categories; the "need-to-know" information and the "nice-to-know" information. When they take the test, they must answer 100% correctly on the "need-to-know" information. If they don't do everything perfectly in the "nice-to-know" category, then they get additional comments and education as to why these issues are important. They learn cardiopulmonary resuscitation, and basic ventilator maintenance; they

spend a great deal of time learning about the developmental needs rehabilitational needs of the child.

I am now going to go through a series of eleven slides to give you a feeling for what is going on in a special care unit for ventilator-dependent children. I emphasize that this is the unit where families are encouraged to be with their children; this is where families are encouraged to accept responsibility for their children. This unit is where the children live. We try as much as possible for the care to be humane. When children first come in, we obtain 48 hours of pulse oximetry and capnograph 1 strip-chart recording to determne the pattern of oxygenation and ventilation. We correlate pulse oximetry and capnography data with blood gas values, and then we donot get any more blood gases. The monitoring of the children is clinical. These children may not have blood gases for six months to a year at a time. We use clinical scales, patterns of respiratory retraction, respiratory rate and noninvasive monitoring. for evaluation.

We had a child recently transferred to us from Georgetown University Hospital. The child was one year of age. Every day during that first year, he had capillary blood gases. That means every day of that child's life, someone took a steel needle and put it into his foot in order to get capillary blood gases to assess oxygenation and CO_2 exchange. I do not think that that is necessary. I think that clinical scales and non-invasive monitoring give you sufficient information to take good care of these children. You do not need scientific precision in controlling oxygen saturation or CO_2 exchange. You need to develop the safe ranges.

This slide shows a fifteen-year-old mother who has obviously learned to take care of her child. She has a sense that she is no longer intimidated by a ventilator nor by a tracheostomy. She can commute to the hospital after she gets out of school and provide care for the child.

This slide illustrates another mother obviously getting satisfaction out of giving her child well-baby care. You can see that the character by the bed is a function of the parents' and the family's enthusiasm for the child. The muchneed not be a strict hospital environment; it has to be as much as possible a home environment.

This slide shows another child with multiple congenital anomaly (multidigition and backbone syndrome.) He was sent to us from another hospital because they thought that he was in the end-stage of this disease that and he was going to die. The request was that we would teach the family how to take care of him at home on the ventilator so he could die at home on the ventilator. We were able to develop a program of pulmonary rehabilitation so that he could be off the ventilator for a period of time.

But quality of life depends upon the functional ability to accept challenges and to meet those challenges. So even though the presumption was that this child was going to die, we saw no reason to treat him like he was going to die. We continued his education in the hospital and made sure that before he went home, he was guaranteed a program of education at home. Because although we knew he would die, we had no idea when he would die. When he first came to us he was six years ago, since that time he has completed high school, an occupational technical school. He has become a talented artist; he does magnificent work. Now that he has graduated from school, he works as a private artist doing portraits on wine glasses. Although has

become more dependent on the ventilator and less mobile, he nevertheless he has lived much longer than anybody anticipated, and his life is good quality.

Again I would emphasize that this is not just an exercise to keep people alive. This is an exercise to give people a good quality of life.

This slide shows a mother who has learned occupational therapy, physical therapy, speech therapy, and sign language. Parents encouraged to actively participate in the occupational therapy and physical therapy programs. For example, mother knows what toys are necessary to enhance certain motor skills.

In the hospital, there are group activities, too. Here are children getting together. In a rhythm band, where each child has an instrument and gets together for group sessions. Socialization is important. The risk of socialization is crosscontamination with bacteria, but this is "the family" in the hospital, and we feel it reasonable for them to be with and socialize with their family. If they go home, their family that they are socializing with is not a family with a lot of gram negative bacterial infections as in the unit The family is cleaner bacteriologically, and that is one of the great advantages of home care.

The hospital environment does not have to be a painful and a depressing place. Even these children know that although they are tied to technology they can have a good quality of life.

This slide illustrates, another child enjoying life. You can see the tracheostomy swiveling here. But it has a very low profile and clings to the chest. So these children can be very active with its two-point swiveling connections. This slide show the tracheostomy swivel referred to a couple of times. It swivels at this point and there is a port for suctioning.

Now I am going to go through a sequence of the slides on the changing of tracheostomy cannula. The emphasis is that these parents are changing the tracheostomy. The parental acceptance of tracheostomy is actually much better than acceptance on the part of professionals. Once the parents have gotten through the psychological barrier of changing the tracheostomy, the parents become very comfortable with it. Generally they are more skilled than many of the doctors since most of the doctors are new and rotating through the units. The parents are looking after one patient week after week. Thus, the parents become very skilled with routine care and with respect to emergency interventions. For examples the parents are trained to use a stethoscope. They are trained to recognize wheezing, to inspect the tracheostomy site, and to look at the child for clinical status.

When the head is in midposition, there is nothing keeping the tracheos rong inplace As these children get older, they get very active; in order to minimize the risk of decannulation, we have developed a special harness. It goes around the neck and under the shoulder to reduce there is less risk of decannulation.

Once the parents have gone through the process of education, and once we have set up the all of the funding, we go out to the house to make sure that the house is safe and that the community support for the child is in place. Then we will move on to transferring the child home.

These are slides show children going home. This is an older child with a battery operated portable ventilator on the back of the wheel chair. In addition, the child has a portable battery operated suction device and, if necessary, portable oxygen, generally in the form of liquid oxygen. Our emphasis again is on ability. We do not want these children to

be restricted to their beds. We do not want them chained to a larger machine. We want them to use portable battery operated ventilators as much as possible. If we send child home with two ventilators, one is usuallya fixed-base ventilator. This slide illustrates the fixed-base ventilator in the child's room. This child also has a portable ventilator for going-off activities. This is the girl who had a tracheobronchial malacia. These slides show the means for independent activity. This is the heat moisture exchanger in the system prevents insufficient humidity and dried secretions. You can appreciate that the scope of activities available to the child at home is much greater than the scope of activities available to the child in the hospital. Although we may have swings in the intensive care unit, I think the swings at home are better.

Feeding patterns are much much better at home. Children with tracheostomies requiring positive pressure ventilation often have a great deal of trouble eating. The patterns of feeding change dramatically once they are at home eating on a regular basis by regular means. Within the social context of the family, the pattern of eating by mouth improves dramatically, and the children generally start to put on weight very quickly.

In summary, let me say that the goals for ventilator assisted children are very straight forward. As previous speakers have said, the primary mission of our care must focus upon the prevention of chronic respiratory failure. We hope that with the judicial use of oxygen, perhaps with high frequency oscillatory ventilation, perhaps with the use of surfactant, certainly with the avoidance of endotracheal tubes and the necrotizing gram negative infections that are part of endotracheal tube, we can prevent the disease that lead many of these children to chronic respiratory failure. But when we have not been able to prevent the disease or where the condition has evolved from the medical support which nave given, we hope to be able to provide medical stability. We hope to be able to provide the children with wellness. We want to move them, whenever it is possible, into an environment that is not restrictive and which involves the patient's development of cognitive, social, and motor skills as well as physical growth and development, not just for the child but also for the family. We want to promote family autonomy and family coping skills.

Thank you very much.

（文責・西村匡司）