Controversies on Management of Congenital Diaphragmatic Hernias

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The principles of management in congenital diaphragmatic hernia (CDH) cover a high degree of controversy that is still going on. During the first 30 years of this century, the treatment for neonates with congenital diaphragmatic hernia was conservative and inevitably the mortality was very high. Many changes have occurred up to now for congenital diaphragmatic hernia patients regarding subgroups of the etiology, pathophysiology, and treatment. Mainly, researches are going on trying to determine the best time for surgery, the most appropriate type of preoperative and postoperative ventilation, and extracorporeal membrane oxygenation (ECMO), the feasibility of fetal surgery, and measures to predict outcome. The basic controversies do not seem to resolve in the near future for congenital diaphragmatic hernias. [Journal of Turgut Özal Medical Center 1997;4(1):123-128]

Key Words: *Congenital diaphragmatic hernia, extracorporeal membrane oxygenation, ECMO*

Konjenital diafragma hernilerinin tedavisinde çelişkiler

Konjenital diafragma hernisinin tedavi prensiplerinde halihazırda çok fazla sayıda çelişkiler vardır. Bu yüzyılın ilk 30 yılında hastalık biliniyor olmasına rağmen tedavisi konservatifti ve doğal olarak ta mortalitesi çok yüksekti. Etyoloji, patofizyoloji ve tedavi açısından günümüze kadar birçok değişiklikler yaşandı. Şu anda devam etmekte olan başlıca çalışmalar en uygun ameliyat zamanlaması, en uygun pre ve postoperatif ventilasyon teknikleri, ekstrakorporeal membran oksijenasyonu , fetal cerrahi uygulamaları ve prognozun müdahaleler öncesinde belirlenmesi çalışmaları şeklinde düşünülebilir. Ancak konjenital diafragma hernisindeki temel çelişkiler yakın bir gelecekte çözülebilecek gibi durmamaktadır. [Turgut Özal Tıp Merkezi Dergisi 1997;4(1):123-128]

Anahtar Kelimeler: *Konjenital diafragma hernisi, ekstrakorporeal membran oksijenasyonu*

The principles of management in CDH cover a high degree of controversy that is still going on. The indicator of this claim is a simple review of the literature on CDH in the last five years (1-6). Many articles are controversial and almost every article is suggesting that their survival rates are fairly satisfactory when compared to the other series.

During the first 30 years of this century, the treatment for neonates with CDH was conservative and inevitably the mortality was very high. CDH of posterolateral region is named after Dr.Vincent Bochdalek from Chek who identified it in a child in 1848. He speculated that the hernia resulted from a posterolateral rupture of the membrane separating the pleuroperitoneal canal into two cavities. Although Bochdalek's understanding of the embryological development of CDH was wrong, this disease carries his name (7). After surgical

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attempts all ended in death of the babies, in 1946, the defect was reported to be repaired successfully in a baby less than 24 hours old (8).

Many changes have occurred up to now for CDH patients regarding subgroups of the etiology, pathophysiology and treatment. All of the subgroups are extremely important, because, pathophysiological principles, for example, if change, will directly have effect on the treatment principles. In fact, CDH was initially thought to be only an anatomical defect of the diaphragm. Pulmonary hypoplasia was identified in 1953 (9). Pulmonary hypertension was recognized in 1971. All of these components are currently recognized in the management of CDH without any controversy. The mortality of these infants is quite high still in our time (10). Many die in utero or postnatally before diagnosis and a significant mortality still is a great problem of pediatric surgeons remaining yet unsolved, and of course a lot of controversies and not a standardized treatment following it.

Mainly, researches are going on trying to determine the best time for surgery (4,6,9,11-19), the most appropriate type of preoperative and postoperative ventilation, and ECMO (1,20-29,31) the feasibility of fetal surgery (32,33-36) and measures to predict outcome (10,17,23,37-40).

The current controversies can be discussed by dividing into two major groups: I-Conventional management, II-Management with ECMO

I- CONTROVERSIES IN CONVENTIONAL MANAGEMENT

Once the diagnosis has been confirmed, care generally centers on stabilizing the infant's respiratory status prior to surgical intervention. But the time stabilization takes differs from center to center, surgeon to surgeon, and case to case (5,11-13). It is a general truth that unstabilized patients, if undertake an operation, do not have a good prognosis. Nakayama et al. have shown that CDH patients, if operated on as very urgent cases, may have a reduced thoracic compliance by increased intraabdominal pressure, interfering with the ventilation (13,41). But how long will the stabilization take? What does stabilization mean? These are matters of controversy in conventional management, even an hour passing intubated and on ventilator preoperatively can be accepted as a stabilization procedure.

Mainly the efforts of preoperative stabilization are reducing the pulmonary hypertension, maintaining an adequate systemic blood pressure, and optimizing the oxygenation. Intensive follow-up of patients is mandatory and Haugen from Norway even advised echocardiographic monitoring to follow pulmonary hypertension (14). Of course, the ultimate goal is to increase the survival. This approach came into light in 1983 and together with growing ECMO, it developed also sometimes challenging, sometimes augmenting it.

A N/G or O/G tube is mandatory; mask and bag ventilation must be avoided. For mild symptoms, supplemental oxygen would be enough, but for severe respiratory symptoms, especially if transport is necessary, endotracheal intubation is required. The general principles for ventilating a CDH patient are: (1) A high respiratory rate, (2) the lowest possible mean airway pressure, (3) a short inspiratory time. These are used for reducing the incidence of air leaks due to overinflation of the lungs. There are studies showing well that pressure alone is not a factor for causing pneumothorax. De Luca U et al (42) have shown that if we can overinflate with a low pressure, it will create a pneumothorax also, not in the hypoplastic only, but also in the healthy lung, there can occur a pneumothorax. One of the things that may help to protect the lungs can be surfactant therapy. Glick et al (20) have reported fair results with surfactant in CDH to increase the compliance and to protect the lungs hypothesizing that CDH lungs are identical to premature, surfactant deficient lungs. Recent work by several authors such as Barefield et al. (43), and Henneberg et al. (44) has revealed another controversial issue on a relatively new agent, nitric oxide. This agent works well in some patient populations and in some others, the results are not satisfactory.

For closure of the diaphragmatic defects, a real controversy exists. Some of the authors never advocate using a prosthetic material and close the diaphragm primarily or very rarely with a muscle flap while some other surgeons use the prosthesis very liberally as Bax et al. have reported the advantages of reconstruction of the dome of the diaphragm, advising to use them in every case with CDH (45). Most of the surgeons use the prosthetic

materials if they believe there is a real indication. In other words they use them if they think the defect cannot be closed if prosthesis is not used. After returning the intestines to the abdomen, in most of the cases the abdominal wall incision can be closed primarily with all layers included. If not suitable, creations of a ventral hernia with only a skin cover or closure with prosthesis are alternative methods of choice.

One of the controversial issues exists on placement of chest tubes. Some of the authors prefer using ipsilateral chest tubes with only underwater seal but without suctioning suggesting that the total lung volume with CDH is often less than what is necessary to fill the thoracic cavity and postoperative attempts to inflate aggressively can cause pneumothorax particularly on the contralateral side; the negative intrathoracic pressure created by chest tubes to suction may also contribute to the overdistension of the ipsilateral side (De Luca et al.) (42). Some authors recommend using chest tubes for both sides, for example Kathryn Anderson suggests that careful manipulation of the suction, applied to both sides, brings the mediastinum to midline. 5 to 7 cm H_20 pressure applied to ipsilateral side and 8 to 10 cm H_2O pressure applied to the contralateral side avoids both lungs from hyperinflation (46). A balanced thoracic drainage system has been suggested by Tyson and associates (47) where a closed system with variable pressure limits is used to allow air to escape when intrathoracic pressure exceeds normal and to enter when pressure falls below normal. Very delayed surgery, spontaneous respiration and no chest tubes is another interesting method that is suggested recently by Wung et al, the air inside the pleural cavity is said to be absorbed slowly, expanding the lung gradually (48).

The main goal of postoperative management is prevention of pulmonary hypertension as in the preoperative period. The ventilation principles are the same as preoperative ones.

II-CONTROVERSIES ON MANAGEMENT WITH ECMO

If the patient cannot be stabilized with conventional management, what can be done? One of the alternatives is "High Frequency Ventilation (HFV)" and the other is "ECMO". One of the highly controversial issues exists here. There are some centers that after conventional therapy fails, if the trial of HFV fails also, ECMO is never tried for salvage. This means that they use HFV as a last resort and as an indicator of the pulmonary insufficiency. If HFV fails, that means for them that lung hypoplasia is very intense, and not compatible with life, they think of no need to try ECMO.

Authors defending HFV or intratracheal pulmonary ventilation (ITPV) (1,49) say that they depend on HFV because it works with a very low mean airway pressure, it does not cause barotrauma, and does not cause pneumothorax , it doesn't need extensive manpower, it is easily available and at the same time it can be used for small prematures without any contraindications. Some of the authors do not believe that HFV is really efficient. They see no benefit in trial of HFV and go directly on ECMO if they think it is necessary.

In fact, one of the hard points is a preoperative ECMO indication spectrum. For the patients that have been well for some time and have deteriorated rapidly, it is easy to define an indication; their lungs have proved to be sufficient at some point of their lives and can be made sufficient again. But what if for the patients that have never been well at even one point of their postnatal time? How can we tell that ECMO will be worth trying? It is an extensive procedure and it must be tried in the patients worth doing it. This is another controversy that gathers attention and many publications but still needs much more effort to be resolved.

Generally it is very hard to compare even the last two decades with present time. For example, the ratio of the patients with CDH that arrived to the operating rooms has changed enormously mainly because of transport facilities. More, many ECMO patients for CDH are referrals or in other words, outborns and for these patients we can say that the survival rate automatically becomes the rate of very worst patients born outside. For these reasons, it is really hard to create some groups and control groups and to obtain a reliable comparison. Most centers using ECMO have had a survival rate of greater than 50% in those patients that more than 80% mortality is anticipated.

For the CDH patients on ECMO, the main causes of mortality can be pulmonary hypoplasia, pulmonary hypertension, brain death,

bronchopulmonary displasia, and hemorrhage (23). Some ECMO survivors have been reported to die 4-8 weeks after ECMO ended. In Dr. Stolar's 3 cases, sufficient pulmonary parenchyma existed and ECMO helped but then pulmonary hypertension recurred because of a very severe pulmonary hypertensive angiopathy (21). As a summary of selection of patients with CDH for ECMO it can be concluded that a high degree of controversy exists. Much effort is spent to find a way to predict outcome.

For the operation aspect on ECMO there is a general agreement that it must last a short time period, dissection must be minimal, and a more liberal use of patches is advised either for diaphragm closure or wound closure or both.

This disease can not be compared with any other disease needing ECMO because CDH patients rely on only one of their lungs; it has a lower survival rate than the other diseases needing ECMO. Survival has not improved even with advanced technologies. Each center's results for conventional management is different and every article mentions about the lack of randomized trials for conventional management and ECMO and Bohn's criteria have not fit to all of the centers (21,37). ECMO can be used in the postoperative period also. Newman and associates in 1990 suggested that all infants with CDH are candidates of ECMO if clinical need arises (24).

After this discussion, one can easily understand that predicting the outcome of infants and determining the potential success of intervention is hard. Both ventilation parameters and anatomic characteristics have been used to predict outcome in CDH. Anatomic features included polyhydramnios, size of defect and position of the stomach etc (34,40), pulmonary parameters have been $PaCO_2$ values, ventilation index, alveolar-arterial oxygen differences and oxygenation index etc (37,50).

There have been no well-estabished criteria and they differ in quality and quantity from center to center. Vacanti et al. (6) after trying both ways of management consecutively, when compared what they have done in the past, found out that delayed repair after stabilization with ECMO is not different in survival from emergency surgery and ECMO if needed afterwards.

Fetal surgery is a very different kind of approach that can not be compared with anything else, aiming to solve the problem from the very beginning and up to now, for the limited number in its own kind, it has really good results (33-36)

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