Summary&Conclusion

Congenital diaphragmatic hernia (CDH) results from failure of complete fusion of the developing fetal diaphragm; a process that normally occurrs between gestational weeks 6–8. The defect may be anterior (Morgagni hernia) or more commonly, posterolaterally (Bochdalek hernia). Migration of abdominal organs into the thoracic cavity can lead to pulmonary hypoplasia, hypertension and respiratory failure at birth.

Current standard management of CDH in newborn babies usually involves initial ventilatory support and supportive care, to allow labile cardiopulmonary physiology to improve. This is followed by surgical reduction of the hernia (usually through an abdominal, laparoscopic or thoracoscopic approach); and repair of the diaphragmatic defect.

The advent of smaller videoscopic instrumentation has allowed the consideration of thoracoscopic repair of CDH repair, which may lead to faster recovery, shorter hospitalization, and less morbidity. Moreover thoracoscopic insufflation of low pressure leads to hernia reduction with no or little surgical manipulation as the positive intrathoracic pressure exerted by insufflated CO2 could push herniated organs into the abdomen.

However, there are theoretical concerns about the adequacy of ventilatory support (because of the partial lung compression that is usually required) and the development of hypercapnia (because of CO2 absorption) during the thoracoscopic procedure. In addition, it is not possible to thoracoscopically repair intestinal malrotation, which is sometimes present concomitantly. Furthermore, concerns about larger incidence of recurrence, longer operative time, less ease of reduction of the hernia, conversion to open procedure; and longer postoperative ventillation and hospital stay still exist.

Attempting repair of a CDH using thoracoscopic techniques is an endeavor that requires careful planning, patient selection, and cooperation between surgeon and anesthesiologist. Given the possibility that a prolonged minimally invasive approach could precipitate complications of respiratory acidosis, pulmonary hypertension, need for ECMO, and even mortality, preoperative selection criteria that would optimize the chances for a successful clinical outcome have been raised and included an intraabdominal stomach position, good preoperative pulmonary function, clinically stable pulmonary hypertension and a diaphragmatic defect that can be repaired primarily.

Thoracoscopic congenital diaphragmatic hernia is performed with the patient under general anaesthesia and in the lateral decubitus position. Between 2 and 4 trocars can be used. Insufflation of CO2 into the pleural space to partially collapse the lung sufficiently to achieve good exposure of the defect and to reduce the herniated viscera within the abdomen. Following the reduction of the herniated content, the diaphragm is repaired using non-absorbable interrupted sutures (for small defects) or patches (if defects are relatively large). Where technically possible, posterolateral diaphragm stitches are passed around the posterolateral ribs and tied extracorporeally. Patients usually require temporary chest drain insertion and ventilatory support.

As minimally invasive approaches continue to gain popularity in the pediatric surgical community, it is incumbent upon surgeons to report not only their successes but also their failures. Acquisition of honest information will allow the pediatric surgical community to learn from each member's experiences and to apply the growing technology appropriately to infants and children.

CDH continues to be a vexing problem for the practitioner caring for these patients. Progress in this field has been hampered greatly by lack of communication and cooperation between major medical centers and by accepting anecdotal observations as dogma.

The initial impression that CDH was a surgical emergency was incorrect, and evidence to that fact was apparent early on. However, progress was slowed by unwillingness to part with established practice, and unwillingness to cooperate in multicenter trials to scientifically ferret out the best therapies. The still widely-held belief that allowing a patient with CDH to develop or maintain a right-to-left shunt would invariably lead to a downward spiral; and this has resulted in far more deaths occurring secondary to ventilator-inflicted lung injury than would ever have occurred from the shunt alone. Even with the emerging evidence that adopting a strategy of permissive hypercapnea has resulted in dramatic improvement in survival in several centers previously practicing hyperventilation, resistance to this concept remains, and many patients are still being treated with outmoded ventilatory techniques.

Furthermore, the lack of a national registry has slowed the development of severity predictors by which prospective randomized trials of novel therapies could be evaluated, and thus none have been done.

Frustration with the CDH problem has led to attempts to deal with the problem in the womb. Although these trials are based on sound biologic principles, because of maternal invasion and the tremendous infrastructure required to perform

fetal interventions, it is unlikely that this avenue will ever be mainstream therapy for the majority of patients.

The time has therefore come for individual institutions to abandon their parochial views, and to acknowledge that the proper management of infants with CDH is unknown. Fledgling efforts such as the CDH study group, which has linked 65 centers to share prospective data in a central registry, is clearly the way of the future. It is in this similar fashion that many childhood cancers were vanquished with coordinated prospective multicenter randomized trials to find the best therapies.

It is also important to understand that CDH is probably more complex than the widely held belief that pulmonary hypoplasia results from simple compression of the lung by herniated intestine would suggest. More likely there is a complex embryologic process which results in not just small lungs, but abnormal small lungs, a maldeveloped diaphragm, and many other anomalies. Consequently, an understanding of the underlying biology of both normal and abnormal fetal lung development will be the key to decreasing the number of infants born with CDH and to improving the survival and lifestyle of those that are.

For the present time, however, the overall survival of CDH in the United States can be significantly increased with therapies that are currently available, by adopting a few simple strategies:

- CDH is a physiological, and not a surgical emergency, and the infants do not belong in an operating room until they have stabilized.
- Infants with CDH should be treated as all other critically ill children and infants are; namely, to provide adequate ventilatory support and oxygen to meet the metabolic demands of the patient and maintain a preductal oxygen saturation of approximately 90%. Close monitoring of mixed venous O2 saturation, as well as bicarbonate and lactate levels, will assure that adequate oxygen is being delivered.
- Because infants with CDH by definition have pulmonary hypoplasia and a fixed number of alveoli, every attempt should be made to preserve all of the alveoli the child was born with. A single pneumothorax requiring a chest tube is clear evidence of barotrauma resulting in a lung less able to perform its function than it was prior to the injury.
- Pulmonary hypertension and right-to left shunting through the patent ductus arteries and foramen ovale are annoying, but are rarely the cause

of death in infants capable of maintaining an adequate preductal saturation. More often the cause of death is an irrational response to the shunt, resulting in increasing ventilator settings, thereby injuring the already hypoplastic lungs.

- ECMO is a proven technology and undoubtedly does salvage some of the most severely affected infants when conventional means are unable to. As such, it should be instituted expeditiously prior to the onset of barotrauma. Furthermore, infants with CDH ought to be delivered at or near a center with experience in ECMO technology.
- The proper place to evaluate therapies such as surfactant, antenatal steroids, high frequency oscillatory ventilation, inhaled nitric oxide, and other potential therapies that develop, is within the framework of a prospective randomized multicentered trial, so that after a brief period of time the technology can be either embraced or discarded and we can move on. In the days where the catchphrase 'evidenced-based medicine' is so prevalent, it is somewhat surprising that virtually everything that is considered current practice with CDH is not based on any evidence beyond individual anecdotal experience.
- CDH survivors are not 'fine.' But they do have predictable pulmonary, gastrointestinal, and nutritional problems, which respond well to careful follow-up and early intervention.

- Finally, it is important to understand that CDH is not necessarily a grim prognosis. Survivals at the most advanced centers are now approaching 90% and higher with the tools available today. Consequently, it seems clear that most infants with CDH can be salvaged with a simple ventilator, a few drugs, a dedicated and experienced team, and not necessarily the newest and trendiest innovation in critical care. The challenge before us all is to review the wreckage of the past four decades where CDH is concerned, and as a group turn our modus operant away from the "quest for the Holy Grail" and toward a rigorous 'evidenced-based' and scientific evaluation of the problem and the proposed solutions.