PERINATAL CARE CONFERENCE 2016

Complex Prenatal Diagnoses:

A Holistic Approach to Management & Treatments

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Complex Prenatal Diagnoses: A Holistic Approach to Management & Treatments

PERINATAL CARE CONFERENCE 2016 took place on Saturday 30th January 2016 in the Royal College of Surgeons, St Stephens Green, Dublin 2, Ireland.

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INTRODUCTION

The Perinatal Care Conference was an entirely unique and important experience which was designed to educate healthcare professionals and medical practitioners from a wide variety of backgrounds - nurses, psychologists, bereavement specialists, palliative care providers, cardiologists, neonatologists, obstetricians and others - in the field of perinatal care. New and groundbreaking developments in this field were also shared.

The role of perinatal care and perinatal hospice is far too often underdeveloped, nationally as well as internationally. With this in mind, the Perinatal Care Conference was held to bring together the diverse array of medical professionals who are brought on board to treat both mother and child when a family is given a difficult diagnosis of a life-limiting condition.

With the advent of ever increasingly sophisticated pre-natal testing technology, parents are now more frequently confronted with findings before birth that can be less than ideal. When faced with a prenatal diagnosis of anencephaly, or Trisomy 13 or Trisomy 18, it is not unreasonable for mothers to expect the same support for themselves and their child - throughout their pregnancy and delivery - as any other patient with a serious condition has the right to expect. These patients deserve our full support and our respect.

At the Perinatal Care Conference, a number of distinguished international speakers, with a tremendous range of expertise, shared their wealth of knowledge and experience. Significantly, we also heard from parents of babies with life-limiting conditions so that we could learn from their experiences and their insights.

This report offers a summary of each of the medical presentations and highlights the key points each of the speakers made. It contains many interesting and important reports on new developments for medical professionals and families alike. The Conference observed that the area of perineonatology has never been so exciting. From babies being born and thriving at ever earlier gestations, to new and revolutionary developments for treating conditions that were previously thought to be invariably fatal, this sub-specialty is opening a whole new world of possibilities.

It is our hope that this conference advances the conversation around improving perinatal care in our country, and demonstrated that we can improve care for babies with life-limiting conditions and for their families.

Yours sincerely,

DR. EAMON P.J. McGUINNESS
MA, FRCPI, FRCOG

Former Chairman of the Institute of Obstetricians & Gynaecologists Ireland

COMFORT AND CARE

Meeting the needs of the baby and the family where a life-limiting condition is diagnosed

"Before death, there is life, and the moment of birth for these babies is a moment of joy," said Dr. Elvira Parravicini, presenting on the importance of comfort care for babies with severe life-limiting conditions, and meeting their specific needs as well as those of their families. This concept is at the heart of perinatal hospice care.

Dr. Parravicini was very clear on her goals as a neonatologist. "My mission is to save babies," she stated. She said that, in the United States, comfort care usually means just letting babies with severe conditions die on their own. When confronted with this reality, Dr. Parravicini felt that, while it might not always be appropriate to provide invasive procedures in such cases, proper care should be given to the baby for the length of his or her natural life. With this in mind, Dr. Parravicini tried to think of ways to make these babies comfortable, and come up with guidelines to provide both standardised and individualised care.

"We want to accompany these babies until their last day of life, being aware that they have dignity," Dr. Parravicini stated, regarding the role of medical staff and the need to create a proper environment for both the babies and their families.

Dr. Parravicini stressed the importance of making the baby comfortable. She noted that being comfortable is more than simply being without pain, and, especially for a baby, can include being fed, spoken to, held, and more. A study from 2000, conducted by Gray et al., found that babies who experience skin-to-skin contact with their mother were observed to have decreased grimacing, crying, and a more stable heart rate when undergoing a heel stick. "It acts almost as an anaesthetic," Dr. Parravicini observed. Additionally, a 2012 study, conducted by Bellieni et al, on multisensorial saturation for infants' pain, found that feeding, touching, and talking to infants decreased their levels of pain.

Based on these principles, Dr. Parravicini and her staff have developed certain guidelines for comfort care so that the basic needs of the babies can be met, and they can be as comfortable and happy as they can for as long as possible.

Guidelines for Comfort Care

- Be welcomed - facilitate infant/family bonding
- Be warm and clean - kangaroo care (skin-to-skin contact), swaddled, and held
- Neither thirst nor hunger - breast, bottle, or syringe fed
- Not suffering pain - assessment and treatment of pain

Dr. Parravicini discussed with the conference attendees the misconception that babies with severe life-limiting conditions suffer due to their conditions. She told the audience that, in her experience

of dealing with more than 200 of these babies, any pain or discomfort was quickly relieved, and these babies tend to slip away - they have a peaceful experience of death.

However, rather than focusing on the end, Dr. Parravicini thinks more people should focus on the life of the child, and making the most of it for them and for their families.

Dr. Parravicini said that while most of these babies will have very short lives, she felt it is important for these patients and their families to focus on the lives they would share, whatever length it would be, and to make them as comfortable and happy as possible. She shared photographs and testimonies of families who had emphasised the importance of having shared time, and who spoke about the opportunity for siblings and extended family to bond with a baby who had not lived for long after birth. According to families, that time was a celebration of the lives of each of these babies, she said. Resources were made available so that families could make handprints and footprints and capture other memories.

Dr. Parravicini emphasised that not one of her patients ever died alone in a bed. They are always being held by a parent, a family member, a doctor or a nurse. They are never left alone and there is never a time when one of their basic needs is not being met.

She also presented some cases which underlined differences in prenatal and postnatal testing, differences which she emphasised should not be described as mistakes, since no-one was at fault. One baby was diagnosed with Trisomy 21 (Down Syndrome) with tetralogy of fallot and lobar holoprosencephaly, and Dr. Parravicini said that the presence of all three indicated that the baby's life might be limited, but when the baby was born, what looked like a lobar holoprosencephaly was actually an arachnoid cyst, something easily treatable. The baby is now 7 years old, and living a full life with Down Syndrome.

Dr. Parravicini also discussed her team's role in following the baby's lead: "If the baby is strong and lives, we feed them, we support them, and then we learn so many things about the nature of these diseases".

In summary, Dr. Parravicini said that personalised comfort care helped parents to love their children when a severe life-limiting diagnosis was made, and that was the motivation and goal of the care she and her staff provided.

See more on the **studies** described here:

Gray L, Watt L, Blass E. Skin-to-skin contact is analgesic in healthy newborns. Pediatrics 2000;105:14–19. doi:10.1542/peds.105.1.e14 [PubMed]

Carlo Valerio Bellieni, Monica Tei, Francesca Coccina, and Giuseppe Buonocore, Sensorial saturation for infants' pain, The Journal Of Maternal-Fetal & Neonatal Medicine Vol. 25, Iss. Sup1, 2012

Dr. Elvira Parravicini is a neonatologist at New York Presbyterian Hospital and a Professor of Pediatrics at Columbia University.

Dr. Parravicini is also a leader in the movement for Perinatal Hospice care and is the founder and medical director of the Neonatal Comfort Care Program at Columbia University Medical Center, which is a model centre for such treatment in the world.

You can watch **Dr. Parravicini's** presentation here: www.perinatalconference2016.com

FIXING HEART DEFECTS BEFORE BIRTH

In-utero treatment of Hypoplastic Left Heart Syndrome

In-utero procedures to fix cardiac defects are steadily moving forward and proving successful in addressing Hypoplastic Left Heart Syndrome, a leading practitioner in foetal cardiac intervention told the conference.

In his presentation, Dr. Wayne Tworetzky, of Boston Children's Hospital and Harvard Medical School, discussed what has been learned from over ten years of research and foetal cardiac intervention, which is slowly leading to the growing use of in-utero treatment of Hypoplastic Left Heart (HLH) Syndrome, a very serious cardiac condition. There is now a real possibility that this procedure might one day be widely available for all unborn babies prenatally diagnosed with this serious condition.

HLH Syndrome is a congenital defect that affects normal blood flow through the heart. As the baby develops during pregnancy, the left side of the heart does not form correctly.

"When an unborn baby has hypoplastic left heart, specifically with aortic valve stenosis, the aortic valve becomes very narrow and the blood flow diminishes causing increased stress on the heart muscle and very serious damage to the heart," Dr. Tworetzky told the conference. "Blood leads away from the left heart, causing the left heart to fail to grow properly."

He observed that for the foetus, it was almost the equivalent to where the blood flow to the heart muscle is being compromised and damage to the myocardium ensues, adding that it was "almost equivalent to the foetus having a heart attack".

Dr Tworetzky told the conference that he had spent time in Dublin in 1998, presenting evidence to the European Paediatric Cardiology conference showing it was possible to stabilise patients with Hypoplastic Left Heart Syndrome where the diagnosis was made before birth. These patients had a significantly improved outcome.

He presented a case in which the unborn child was progressing toward HLH Syndrome and underwent a procedure using the same balloon used in coronary artery angioplasty, as the foetal heart valve is about the same size as an adult coronary artery. "So we use the same balloon, we inflate the balloon and open up the valve. In this particular case, the patient was lucky enough, the function recovered significantly, and this baby, now at 6 or 7 years of age, has a very well-functioning left ventricle, and we really think that we averted progression towards HLH Syndrome and in this particular case we managed to get there just in time."

Dr Tworetzky cautioned that this did not happen in every case, and also presented a case where the procedure had gone perfectly but the heart did not respond. The treatments available have slowly improved over the years but this condition remains a very serious heart defect. He referred to a 2014 study which showed that, in at least half the patients who underwent foetal aortic valvuloplasty, progression to HLH Syndrome was averted. This is significant because a third of babies who are born with HLH Syndrome do not survive beyond 3 or 4 years.

The field of paediatric cardiology had seen considerable progression he said, and many serious heart defects were being treated, but there were still many challenges including development of treatments for the preservation of the myocardium.

Dr. Tworetzky pointed out that the current challenge is to move from simply studying the diagnosis to working towards a practical therapy and treatment. He said that doctors need to ask:

- How can we work to improve the lives of the babies who are affected by this very difficult diagnosis?
- What are the most viable treatment options?

There are several accepted foetal therapies for the syndrome at the moment. Currently, most of these therapies focus on optimising heart function, but few treat potentially lethal conditions. The aim of foetal cardiac interventions is to promote ventricular growth and function and to improve survival. Dr. Tworetzky discussed how these interventions (which use an ultrasound-guided needle to fix the left heart) are still in their infancy and therefore only a few have been performed, and with each intervention they become more successful and precise, with the rate of technical successes rising steadily.

In the longer term, these patients will need heart transplants, but the good news for these children and for their families is that they are living beyond infancy. The procedure can be performed as early as 19-20 weeks gestation, Dr Tworetzky said.

The science behind these in-utero operations is slowly but steadily moving forward. This is good news for parents who are given news before birth that their child has a serious heart defect.

Soon it will be standard procedure to fix cardiac defects in-utero. These procedures are becoming more successful and more patients are being born without HLH Syndrome and with two working ventricles.

See more on the **study** described here:
Freud LR, McElhinney DB, Marshall AC, Marx GR, Friedman KG, del Nido PJ, et al. Fetal aortic valvuloplasty for evolving hypoplastic left heart syndrome-postnatal outcomes of the first 100 patients. Pub Med. Circulation. 2014;130:638–45

DR. WAYNE TWORETZKY is the Director of Foetal Cardiology in the Department of Cardiology and the Advanced Foetal Care Center at Boston Children's Hospital, an Attending Physician in Cardiology at Boston Children's Hospital, and an Associate Professor of Pediatrics at Harvard Medical School.

In addition, he is the founding member of the newly formed Foetal Heart Society and associated multi-center Foetal Cardiology Research Network. Boston Children's Hospital takes an innovative approach to treating HLHS, was ranked number one for heart care and heart surgery in 2010, and is the world's largest and most experienced programme for foetal cardiac intervention.

You can watch **Dr. Tworetzky's** presentation here: www.perinatalconference2016.com

SERIAL AMNIOINFUSION AS A POTENTIAL TREATMENT FOR BILATERAL RENAL AGENESIS

Dr. Meredith Birsner was part of a team at Johns Hopkins Hospital who became the first to successfully treat a baby in-utero for Bilateral Renal Agenesis, or Potter's Sequence. Their patient is the first child to survive, and now thrive, after a prenatal diagnosis of Potter's Sequence.

Dr. Birsner presented on the case, which had been widely reported in the American media, to a raptly attentive audience at the Perinatal Conference.

Dr. Birsner told the audience that the pregnancy had seemed unremarkable at the initial stage. It was not until the abnormality scan took place at about 20 weeks that it appeared the baby lacked kidneys, stomach, and seemed to have clubbed extremities.

Further scans seemed to show an abnormal head shape, hypo-plastic lungs, abnormal chest shape, bilateral clubbed feet, and the bladder and stomach were not visualised. These serious issues are consistent with Bilateral Renal Agenesis, or Potter's Sequence.

With Potter's Sequence, due to the lack of kidneys, a lack of amniotic fluid arises, which leads to severely impaired development, including underdeveloped lungs and other characteristics. This can result in very serious harm to the baby.

Dr. Birsner informed the conference that the parents in the case were offered a termination three times but they repeatedly refused. During a third consultation, at Johns Hopkins Hospital in Baltimore, the mother of the baby initiated the option of amnioinfusion.

Amnioinfusion was offered as an experimental option to the mother after lengthy discussion with the medical teams, and the risks to both herself and her baby were explained to her. This was a procedure that had not been carried out for this purpose but it was hypothesised that there was potential for this sort of therapy to succeed.

The unborn patient's mother underwent the groundbreaking procedure, overseen by Dr. Birsner and her colleagues. Saline solution was injected into her womb five times before the premature birth of her daughter between 24 and 28 weeks gestation, when the mother went into labour and the baby was delivered.

After the first amnioinfusion, the patient came back 5 days later for a follow-up ultrasound. Doctors saw a rise to subjectively normal amniotic fluid levels, a decrease in the foetal heart size to the upper limit of normal, and they observed that the thoracic to abdominal ratio had also normalised. Foetal breathing movements and the presence of the stomach were also noted. Four further amnioinfusions were performed.

The baby girl was born prematurely at 28 weeks, and despite lack of kidneys, the neonatologist noted excellent respiratory effort, and no intubation was required. Respiratory distress syndrome was noted on the chest x-ray; however, there were no visible stigmata of the Potter syndrome or sequence. She spent 19 weeks in the NICU and was then able to go home with her parents with the aid of regular dialysis treatment.

The little girl experienced some very mild developmental delays but is now doing very well and hitting significant milestones. At the time of the presentation, the patient was preparing for a kidney transplant with her father acting as the donor (she has since undergone the transplant, which was successful).

Following the success of the procedure, Dr. Birsner and her colleagues were inundated with requests for serial amnioinfusion therapy. As a result, they have developed a set of criteria that prospective patients must meet in order to be considered for this developing therapy.

Inclusion criteria stipulate that the baby must be a singleton foetus between 16 and 24 weeks with Bilateral Renal Agenesis and an Amniotic Fluid Index (AFI) of less than 2. Parents must be willing to stay in the Baltimore area and deliver at the Johns Hopkins Hospital.

In terms of exclusion criteria, they considered any concurrent major congenital anomalies except those that could be directly attributed to anhydramnios as well as renal agenesis. Additional exclusion criteria include spontaneous rupture of membranes and aneuploidy. Additionally, they require that parents go through procedure counselling with a genetic counsellor and a neonatologist. Parents must be made aware of all of the risks associated with the procedure including risks of maternal and foetal death.

Dr. Birsner told the conference that an analysis of the cost-effectiveness of the therapy had been carried out by Dr. Jessica Bienstock and Dr. Erika Werner of Brown University, and the analysis had found that there are plausible conditions under which serial amnioinfusion is cost-effective for bilateral renal agenesis. Time on dialysis, probability of survival to transplant, overall life expectancy and quality of life after transplant significantly affect whether this intervention is cost-effective.

In relation to the future for renal agenesis, Dr. Birsner told the Conference that there have since been two as-yet-unpublished cases successfully treated with serial amnioinfusion in the United States, while others have not been successful.

Regarding this successful case, Dr. Birsner reported, “This child has been an enormous source of joy to this family.”

See more on the **study** described here:
Bienstock J.L., Birsner M.L., Coleman F., Hueppchen N.A. Successful in utero intervention for bilateral renal agenesis. Obstet Gynecol. 2014;124(2 Pt 2):413–415

DR. MEREDITH BIRSNER is an Assistant Professor of Maternal-Foetal Medicine (High Risk) at Thomas Jefferson University in Philadelphia, Pennsylvania, USA.

She completed her residency in obstetrics and gynaecology and fellowship in maternal-foetal medicine at The Johns Hopkins University in Baltimore, Maryland.

You can watch **Dr. Birsner’s** presentation here: www.perinatalconference2016.com


SPEAKER


SPEAKER

PERINATAL CONFERENCE 2016

EVERY CASE IS UNIQUE

Delivering optimal medical care to babies born with Trisomy 13 and Trisomy 18

Doctors need to support families where a prenatal diagnosis of a chromosomal anomaly is made, and be sensitive to their situation, using the right words and the correct information, according to neonatologist Dr. Martin McCaffrey.

These are babies whose lives may be very short, and Dr McCaffrey said he had witnessed negative attitudes all too frequently, both from medical practitioners and in the existing medical literature, toward babies prenatally diagnosed with Trisomy 13 and 18.

“This is what I was raised with in medical school: that these are babies who don’t survive. That these are babies who have fatal anomalies, lethal anomalies……We were doing families a great favour, by the way, by trying to expedite the demise of their baby,” he said.

Dr. McCaffrey expressed regret that medical professionals carry their own preconceived biases (often left over from medical school and outdated medical textbooks) and that this bias can have a deep and lasting impact on the way society looks at these babies, and the way expectant parents look at their own babies who receive a complex prenatal diagnosis.

He said that much of the data being used with regard to survival for babies with Trisomy 13 and 18 was outdated, while large studies sometimes did not include details of interventions or care after birth for these babies. Pointing to a major 2015 study by RE Meyer et al, Dr McCaffrey noted that this study found survival among children with T13 and T18 to be somewhat higher than those previously reported in the literature, consistent with recent studies reporting improved survival following more aggressive medical intervention for these children. He observed that the data showed that babies who made it to 28 days had a significant chance of further survival, with 38% of those who survived to 28 days reaching at least the age of 5 years.

The language doctors use is crucially important both in the medical literature and when they speak to parents, he told the conference. The language medical staff use at these critical junctures can inadvertently pressure parents to terminate or not to resuscitate.

Dr. McCaffrey was clear. "I'm not saying that doctors must intervene and perform surgeries on every baby. What I am talking about is offering basic care and considering interventions like breathing tubes, for babies who need them."

Dr. McCaffrey pointed to a study which asked an extensive social media network of parents whose babies were diagnosed with Trisomy 13 and 18 about their interactions with doctors. What he discovered shocked him; some 61% of parents reported feeling pressured to abort their babies after the diagnosis was made.

Additionally, some parents felt that the manner in which doctors spoke to them at the time of diagnosis was insensitive, unprofessional, and dehumanising towards their babies. Parents reported that:

- "I was told by the geneticist that the only way I could make an appointment with the main obstetrician was if I was booking a termination."
- "An OB asked me if I wanted to burden my other kids with such a sibling. Another doctor (Ob/Gyn) told me these children lacked the will to live, that they were without a soul, unable to experience being alive."
- "After we confirmed, again, that we would not terminate, we got told that 'the best thing that can happen now is if your baby dies then you can get over this and try again.'"
- "We found out when Addalyn was 2 days old she had T18 and they told us they wouldn't judge us if we chose not to feed her."
- "A paediatric cardiologist said to me, 'You need to terminate. If you don't, I won't touch her when she is born.'"

Dr. McCaffrey emphasised again the importance of sensitivity, and of providing factual and useful information to parents in these situations. He pointed out that some medical practitioners had no experience with these conditions, and, because the education they had received on the conditions is outdated, they may inadvertently mislead parents as to outcomes for children with Trisomy 13 and 18. Doctors may also be under the false impression that mental impairment equals suffering, which is not true.

Doctors need to support families with a severe Trisomy prenatal diagnosis, he added. Medical staff need to work with families to provide a birth plan and support plan that best meets their needs. They need to discuss options for parents who are meeting resistance from facilities to support live birth or provide medical care after birth.

Dr. McCaffrey pointed out again that language is of paramount importance.

"Words mean everything. Language is everything," said Dr. McCaffrey. " At the end of the day what we are fighting for are these wonderful, beautiful children who may have very short, limited lives."

See more on the **studies** described here:

Meyer RE, Liu G, Gilboa SM, Ethen MK, Aylsworth AS, Powell CM, Flood TJ, Mai CT, Wang Y, Canfield MA, for the National Birth Defects Prevention Network. 2016. Survival of children with trisomy 13 and trisomy 18: A multi-state population-based study. Am J Med Genet Part A, published online 2015, doi: 10.1002/ajmg.a.37495,
Guon, J., Wilfond, B., Farlow, B., Brazg, T., Janvier, A. Our children are not a diagnosis: The experience of parents who continue their pregnancy after a prenatal diagnosis of Trisomy 13 or 18. Am J Med Genet A. 2014;164A:308–318.

DR. MARTIN McCAFFREY is a Professor of Paediatrics at the University of North Carolina Chapel Hill and a Board Certified Neonatologist. He served as Chief of Neonatology at Naval Medical Center San Diego, the Navy Surgeon General's Consultant for Neonatology, and Chairman of the Navy Perinatal Advisory Board.

He is the Director of the Perinatal Quality Collaborative of North Carolina and serves on the National Quality Forum and Joint Commission Perinatal Steering Committee.

You can watch **Dr. McCaffrey's** presentation here: www.perinatalconference2016.com

WHEN THERE IS NO CURE

Training neonatal staff for the future of neonatal palliative care

Dr. Paula Kelly is a Clinical Nurse Specialist in oncology outreach and palliative care at the Louis Dundas Centre for children's palliative care in London. This service supports babies, children and young people with palliative care needs and their families in hospital, home, and hospice settings in London and the South East of England.

In her presentation, Dr. Kelly spoke about the important and often under-discussed issue of training neonatal staff for palliative care cases. Very often medical staff - even those used to caring for very sick babies every day - are nervous at the idea of caring for children with such severe conditions as anencephaly and Trisomy 13 or 18.

Dr. Kelly and her team recognise this as a problem as more children die in the perinatal period than in any other period of childhood, yet only 24% of critically ill perinatal patients are referred to palliative care. Despite these facts, there are multiple studies showing that parents of these patients report being dissatisfied with their care options.

This is a major problem as both the neonatal patients with life-limiting conditions and their families are in need of support from those who are confident and properly equipped to provide it. There are many specific issues faced by this population. There are serious social implications to the pregnancy. Parents may feel pressured to terminate by family and friends if they are given a severe prenatal diagnosis.

Dr. Kelly also reported that with this population, there is a very high level of diagnostic uncertainty, and due to the special circumstances of each of these patients, who have a wide range of conditions and needs (and whose families require different sorts of support), it is even more necessary for specialised neonatal staff to focus on these patients and their families.

Dr. Kelly proposed to the conference that it is necessary to establish specific education programmes for this population rather than add on to existing palliative care education programmes. Besides the dissatisfaction noted by parents of patients, Dr. Kelly and her staff also felt that the low rate of referrals for palliative care was a reason to institute very specific educational initiatives.

Dr. Kelly and her staff have undertaken a neonatal palliative care educational initiative in order to try and facilitate and improve earlier access to palliative care services for babies and families in the London area, and they would like to see that effort grow.

As part of their efforts to bring this educational outreach to as many neonatal medical staffers as possible, the programme was launched in 22 neonatal units across the wider London area. Each intervention consisted of a four-hour-long workshop. As there is limited research into the importance of education in neonatal palliative care, surveys were passed out both before and after these workshops.

Dr. Kelly and her team received 200 survey matches pre and post workshop. One of the biggest

issues they identified from neonatal staffers (who were mostly nurses), when dealing with cases of life-limiting conditions, was that of confidence. Medics were most confident about collecting mementos and were also confident discussing the signs and symptoms of the baby's condition.

However, less than half were confident in discussing care for a dying baby in a setting outside the NICU, either in a hospice setting or at home. Other areas where they lacked confidence were discussing sibling concerns, and how to help siblings deal with the short life of the baby.

Dr. Kelly discussed the reasons that neonatal staff gave which made them less likely to make a referral to a palliative care service for a baby with a life-limiting condition. Many felt that the prenatal prognosis was uncertain, and they were uncertain about approaching parents with the option of palliative care when they didn't know the outcome themselves. Additionally, many felt that they were diminishing parental hope by referring to perinatal palliative care.

However, Dr. Kelly reported that following the workshop, practitioners felt more confident about their knowledge and about making a referral. Attitudes towards palliative care had improved significantly. Following the workshop, symptom management was no longer seen as the main goal to be achieved, and the fear of diminising the hope of parents was also reduced.

Dr. Kelly found that after only a short educational intervention, neonatal staff were more confident about making referrals for palliative care and attitudes were more in line with the goals of perinatal palliative care - which are, ultimately, to provide the best care possible to neonatal patients with severe life-limiting conditions, and to provide the support that their families need.

See more on the **studies** described here:

Twamley K, Kelly P, Moss R, Mancini A, Craig C, Koh M, Polonsky R, Bluebond-Langner M. *Palliative Care Education in Neonatal Units: Impact on Knowledge and Attitudes (2013)*. BMJ Supportive and Palliative Care 3:213-220 doi:10.1136/bmjspcare-2012-000336.

Mancini A, Kelly P, Bluebond-Langner M (2013) *Training Neonatal Staff for the Future in Neonatal Palliative Care.* Seminars in Foetal & Neonatal Medicine 18: 111-115.

DR. PAULA KELLY is a registered children's and general nurse. She has a doctorate in Medical Sociology from Queen Mary's College, University of London, and a Masters degree in Anthropology from Brunel University. As an experienced children's nurse, the majority of her clinical work has been in caring for children with life-threatening and life-limiting conditions in the home setting. She joined the Louis Dundas Centre for children's palliative care as a Clinical Nurse Specialist in oncology outreach and palliative care in June 2015.

You can watch **Dr. Kelly's** presentation here: www.perinatalconference2016.com

EXAMINING THE EXPERIENCE OF PARENTS

The experience of families with children with Trisomy 13 or 18

The experiences of families with children with Trisomy 13 and 18 are overwhelmingly positive, and offer a sharp contrast to the attitudes and language used by medical practitioners in prenatal diagnosis, medical researcher and parent Barbara Farlow told the conference.

Ms. Farlow's published research into the experiences of families with children with Trisomy 13 and 18 was inspired by her own experiences following the birth of her daughter Annie, who was prenatally diagnosed with Trisomy 13 at 21 weeks gestation.

Ms. Farlow related the very difficult circumstances following her daughter's birth and death in 2005. Annie was born full-term and was mildly afflicted. Her only serious issues were hypoglycemia and an undiagnosed airway problem. The Farlows spent the first six weeks in the hospital with Annie, and a family member was with her at all times. To her family's relief and joy, she was eventually able to go home with her parents. Her siblings greeted her with a family celebration including chalked messages which said 'you rock this house' and 'you're kicking butt'.

However, Ms. Farlow reported, they were eventually advised to return to the hospital as a conservative measure when Annie began to experience some respiratory distress. Within 24 hours of returning to the hospital, Annie died suddenly at 80 days. Ms. Farlow and her husband later found out that Annie was placed under a Do Not Resuscitate (DNR) without their permission. The medical care Annie received when she died was deemed by the Coroner's Paediatric Death Review Committee to be "inappropriate."

One of the main problems the Farlows faced was the medical environment into which Annie was born. That year, the health ministry of Canada launched a co-ordinated prenatal genetics programme. Rather than seeking ways to better support babies like Annie and families like the Farlows, it seemed to promote the stigma of babies with severe Trisomy. Providers taking part in the programme were shown a Powerpoint presentation showing a stillborn baby with cyclopia (a very extreme form of Trisomy 13) next to a picture of a mythical Cyclops, essentially dehumanising a very real child.

Ms. Farlow discussed how, at the same time, a policy analysis was published for expanding screening for babies with Down Syndrome, showing a cost-saving incentive for prenatal genetic screening. "The standard of care for children such as Annie," Ms. Farlow told the audience, "remains sadly low across the board." At one hospital, the standard care procedure for children born with Trisomy 13 was to simply administer morphine every three hours until the death of the infant.

Ms. Farlow related how she was spurred on by her own experiences to conduct a literature review, and found that reports written about Trisomy in medical journals were negative, referencing one article in the Lancet in 1992, which said that perhaps babies with Trisomy 18 should be helped to die. Further study led her to connect with other parents via social networks and she found that the experience of parents of children with Trisomy could be very different to what was portrayed in medical publications.

Collaboration with Drs Annie Janvier and Benjamin Wilfond led to the publication of an important paper, The Experience of Families with Children with Trisomy 13 and 18 in Social Networks (Pediatrics, 2012), which looked at the responses obtained in 372 parent surveys. The research found that medical practitioners had been overwhelmingly negative when relaying results indicating Trisomy 13 or 18 to parents after prenatal diagnosis. The responses parents recalled included practitioners' statements that:

- The baby was incompatible with life (87%)
- It was a lethal condition (87%)
- The baby would be in constant pain (57%)
- The baby would be a vegetable (50%)
- The baby would have a life of suffering (57%)
- The baby would have a meaningless life (50%)
- The baby would ruin the family (23%)
- The baby would ruin the parents' marriage (23%)

In sharp contrast, parents reported their own experiences in a very different light. In fact, the study found that 98% of parents reported the overall effect of their child's life was positive, regardless of longevity. They overwhelmingly reported that their babies born with Trisomy 13 or 18 were happy and had, in fact, enriched their lives and the lives of their other children. Parents reported a positive, transformative experience even if the baby's life was short.

As Ms. Farlow noted, nobody had asked parents about their experiences and published the findings - instead the clinical guidelines were based on assumptions.

Ms. Farlow said that the study found that a prenatal diagnosis, not the actual anomalies a child might have, was the most significant factor leading to early death. She also noted that over half of parents who were given a prenatal diagnosis reported being pressured to terminate their pregnancy.

Ms. Farlow concluded by saying that positive changes were occurring, and that she concurred with the World Health Organisation's recommendations, which state that palliative care should be offered at the beginning of life-limiting diagnosis and at the same time as any possible curative efforts, recognising that the transition to purely palliative or hospice care may occur later.

BARBARA FARLOW is an engineer who was redirected in her life to become a researcher after the death of her daughter Annie who had Trisomy 13.

Barbara realised that physicians don't understand the family experience of giving birth to a child with a life-limiting condition. As a result, she partnered with two physicians who are leaders in paediatric ethics to survey and publish the lived experience of over 300 parents who gave birth to a child with Trisomy 13 or 18. She is a co-founder of the International Trisomy 13/18 Alliance.

You can watch **Ms. Farlow's** presentation here: www.perinatalconference2016.com

HEALING AFTER LOSS

Bereavement care following perinatal loss in a large Dublin maternity service

Bríd Shine addressed the Conference in relation to the current care practices in Perinatal Palliative Care and Bereavement in the large tertiary referral centre where she works as Bereavement Midwife at the Coombe Hospital.

Ms Shine began by acknowledging all other speakers at the conference and outlined the huge impact perinatal death has on the lives of bereaved parents and their families. She discussed how its nature is often sudden and unexpected and how it results in a profound shock and sadness, and she said the death of a baby has been defined in the literature as a major bereavement and some families may even experience it as a disenfranchised grief. She talked about the unique nature of grieving the loss of a baby and referred to it as prospective grieving.

Ms Shine sought to clarify that enhancing Perinatal Palliative Care did not involve building a perinatal hospice, but rather, it involves bringing the "ethos" of hospice into our maternity care services in Ireland. She cited the Irish Hospice Foundation's (IHF) role in enhancing End of Life Care in Ireland and how the Coombe Hospital was one of the first maternity units to endorse the IHF's "Hospice Friendly Hospitals" programme. She clarified that what was referred to in the U.S. as Perinatal Hospice is referred to in Ireland and the UK as Perinatal Palliative Care, as its broad interdisciplinary team approach with family-centred care is the same.

Ms Shine reflected on Ireland's rich history in caring for the dying, and drew from Celtic philosophy to highlight old care practices that understood and respected the enormous transformative events that are felt at birth and death. She also outlined Ireland's diverse cultural population and the different cultural needs around care of the dying.

Ms Shine went on to highlight the more painful history of how Ireland responded to perinatal loss in the last century and recent decades. She talked about the lack of compassionate care that some families experienced in the past, as a result of health care staff's lack of knowledge and understanding of the impact of the death of a baby, and the care practices now known to afford healing to bereft families.

In acknowledging the painful past of perinatal bereavement care, Ms Shine talked about the importance of offering all families in maternity care services compassionate and non-judgemental care, highlighting our collective need to be respectful of each individual family's decision-making process.

She credited the voices of bereaved parents as being influential in changing care practices and how they have played and continue to play a huge part in "turning the tide" in perinatal bereavement care in Ireland today. Ms Shine outlined the current contemporary grief theories that have helped to shape and change our understanding of the grief process, citing memory-making as being linked to the Continuing Bonds philosophy - making care evidence-based practice.

Ms Shine outlined the wide interdisciplinary teams involved in Perinatal Palliative Care where she

works. She outlined those key disciplines in maternity hospitals as well as the key Primary Health Care teams in the community. This included mention of the specialist perinatal palliative care services for families who may have the opportunity of bringing their baby with a life-limiting condition home. She presented case examples throughout and gave the statistics of foetal anomalies diagnosed in the Coombe Hospital for 2015.

Ms Shine talked about the power of presence when companioning with families facing the anticipated death of their baby and the effective tools required to facilitate transformation of suffering into meaningful living. She outlined what it is like being midwife to the bereaved and the repeated human suffering witnessed in all categories of perinatal loss. She talked about hope as a vital component of bereavement care, and how deep human connections unfold in this area of work.

Finally, Ms Shine drew brief attention to the importance of compassion for Health Care Providers and to research that supports the need for mindfulness practices to be utilised in staff training to prevent stress, burnout and empathy fatigue as staff are impacted in the suffering they bear witness to.

She concluded with a beautiful poem entitled “Our Holly” written by bereaved parents Colm & Laura, with their kind permission, following the loss of their baby Holly to Trisomy 18 (Edwards’ syndrome).

BRÍD SHINE is a registered Nurse and Midwife. She has a higher diploma in Public Health Studies from UCD, and a Masters Degree in Bereavement Studies from RCSI. Bríd is trained in spiritual care of the dying and has certified as a Mindfulness Teacher with the IMA in Europe. With over 25 years’ experience, she has cared for clients in both the hospital and community setting at all stages of life. Bríd’s current post since 2010 is Clinical Midwife Specialist in Bereavement at the Coombe Women & Infants University Hospital.

THE GIFT OF TIME

Providing families with a comfortable and compassionate environment to spend time with their newborn

"We want to empower them to care for their babies," Frances McCarthy, the Clinical Care Coordinator of the Neonatal Comfort Care Program at New York Presbyterian Children's Hospital, emphasised during her presentation on the unique aspects and requirements of a comfort care programme when the baby is not expected to live for long after birth.

Ms. McCarthy began by explaining that neonatal palliative care, or comfort care, is a medical and nursing course of treatment for a child with a severe life-limiting condition. It is not about giving up hope for the child, but simply changing goals and expectations. The goal the medical team is working towards is comfort when cure is no longer a viable option. The idea of comfort, which includes warmth, hydration, human contact, and symptom management, is very important and can be extrapolated to any population.

The characteristics of a neonatal comfort care programme are fairly straightforward, and include a family-centred approach, bringing the family into all aspects of patient care. The programme employs a multidisciplinary team, including physicians, nurses, social workers, and pastoral care. None of these team members can take on this overwhelming job without the others. There must be open communication between parents and team members. It is crucial for the entire staff to be on the same page with regard to the message the family receives. Finally, there must be psychological, emotional, and spiritual support for family and staff. This also helps them to meet the needs of the babies.

Ms. McCarthy noted that when dealing with patients who are not likely to live very long, it is important for the medical team to come to terms with their own attitudes about death and dying, as these are the situations they would be addressing every day. Additionally, they must address and respect the families' attitudes regarding death. Doctors and nurses may experience that people who profess not to be religious at all find that religious traditions are important to them when they experience the death of their child.

In any neonatal comfort care unit, Ms. McCarthy stated, it is the medical staff who set the tone that parents and families follow. Staff need to be calm, supportive and present. The parents will follow the cues of medical staff, and if the staff panic, parents will panic.

She told the crowd that, regardless of whether the baby is in the NICU or on the post-partum floor, it is necessary to provide routine post-partum care. Neonatal comfort care is partly about recognising the needs of the parents, and the parents of children with Trisomy 13 or anencephaly or other severe conditions need to feel like parents for as long as they have their babies.

"We want to empower them to care for their babies," Ms. McCarthy said. This involves teaching parents to care for their children as much as they are able to and for however long they can. She emphasised how feeding one's baby is mothering, as is changing and bathing one's baby. If possible, she and her staff try to give parents the opportunity to do these things. Ms. McCarthy stated that if the child is unable to be fed via the breast or the bottle, they will often teach mothers and

fathers to do syringe feeds so that they can feed their babies themselves.

Ms. McCarthy related a story of one girl born with Trisomy 18 who, after some initial difficulty, was able to latch on and breastfeed without any difficulty at all. She lived for only 12 hours after birth. However, Ms. McCarthy related to the conference, "I promise you, that mother's memory of that baby latching on will sustain her forever. She will never forget that. And she will be forever grateful she got that opportunity."

Ms. McCarthy strongly emphasised that the way the staff interact with parents, both before and after birth, is vitally important and in some ways goes against the grain of what some would expect when confronting a dire diagnosis.

She encouraged medical practitioners to congratulate parents. Regardless of their child's condition, they are still expecting a baby and the relationship between mother and child is profound and important. She encourages staff to ask if the child has been named, and to always use their name. Try to anticipate the family's needs; water, camera, privacy, family, clergy, tissues, etc, and Ms. McCarthy referenced the important work of *Now I Lay Me Down to Sleep* in providing photographers for parents at this time.

After the child is born, Ms. McCarthy explained that it is important to keep parents informed of what is going on and prepare them for what they are going to be seeing and dealing with. She advised, "Let parents know some of the physiological changes they will be witnessing in their babies, and that what they are seeing is a normal part of the process for their children's lives."

Parents should be prepared for changes in their baby's breathing patterns, changes in colour, decrease in movement, decrease in body temperature, gasping, etc. Ms. McCarthy advised doctors and nurses to describe obvious anomalies before parents see the baby but to also comment on the normal and perfect features.

The features of the neonatal comfort care unit are straightforward, but they serve so many people at one of the most difficult times in their lives. By providing basic care to these babies and empowering parents to be parents for as long as they can, they are giving families something truly priceless.

FRANCES McCARTHY is the Clinical Co-ordinator of the Neonatal Comfort Care Program at New York Presbyterian/ Morgan Stanley Children's Hospital (CHONY). Frances was a bedside neonatal nurse at CHONY for 28 years before assuming her position in the Comfort Care Program. She received a Master of Science in Nursing from Columbia University School of Nursing as a Psychiatric Nurse Practitioner and is certified in Perinatal Loss Care. Frances is a member of the CHONY Bereavement Committee and the CHONY Friends of PACT. She is keen to teach new neonatal nurses and others about bereavement support and Perinatal Hospice care pathways.

You can watch **Ms. McCarthy's** presentation here: www.perinatalconference2016.com

OUR BABIES ENRICHED OUR LIVES

"Isabella's lifetime was just 54 days. That doesn't make her any less important."

One of the most meaningful contributions to the Perinatal Care Conference was the participation of parents whose children were diagnosed with severe life-limiting conditions and who came to share their experiences with the conference attendees.

DAN HALEY AND JENNA GASSEW movingly recounted their experience following the devastating prenatal diagnosis of their son Shane with anencephaly in 2014. Dan and Jenna explained how, following their diagnosis, they decided that, though their son might not live long after birth, they would celebrate his life and make the most of their time together for as long as they possibly could.

Before Shane's birth, Dan and Jenna completed a "bucket list" of things to do with their son before birth, which garnered international media attention and a million followers on Facebook. Jenna recalled, "We brought him to the top of the Empire State Building, we felt him kick as we looked out over New York."

Dan and Jenna gave a moving presentation on their experience of going to term with Shane, and stressed the vital importance of perinatal hospice for families with children with life-limiting conditions. "They ask parents what they need, they showed us that they would take care of us and of our son. I can assure you - our son was beautiful. He was my son. The diagnosis didn't change that," Dan told the conference.

SARAH NUGENT, of Every Life Counts - an Irish support and advocacy group for parents of children diagnosed with life-limiting conditions - related her experience with her daughter, Isabella, who lived for just 54 days after birth in 2014 due to a peroxisomal disorder.

Sarah described her daughter as "more beautiful than I could have ever imagined." She said that having Isabella continually referred to as "incompatible with life" was very upsetting. Sarah stated, "I can't begin to explain the fear those words instill into you. I knew she was seriously sick but 'incompatible with life' gave me no information as to what was wrong with her."

Sarah emphasised that parents needed time to parent: "We need a private space, one that allows parents to parent, our families to meet our child. Without costing millions, how can we make small changes to the way we work with families like mine? Perinatal hospice care would provide this model of care, focusing on the value of that baby's place in our family and acknowledging the huge hole they will leave behind." She added, "You do get a lifetime with her - all our lifetimes are different, and Isabella's lifetime was just 54 days. That doesn't make her any less important."

JACINTA MURPHY'S baby Sarah lived for 24 hours when she was born with Trisomy 13; Jacinta recalled that "the day Sarah died, a part of me died also".

Jacinta went on to found Féileacáin, the Stillbirth and Neonatal Death Association of Ireland, in response to "the lack of appropriate services for bereaved parents and families". They run an extensive suite of services, including pre-birth support, psychotherapy and counselling, work-

shops, social work services and play therapy. Their memory boxes and cuddle cots are now available in Irish maternity units, and have proved hugely helpful to bereaved parents and families.

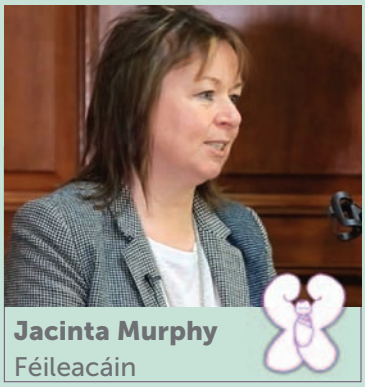
ÍDE O'SULLIVAN, of SOFT Ireland, and mother to baby Lucy, spoke about the need for support both before and after birth, and before and after baby passes away. "Our little ray of sunshine gave us 9 beautiful weeks at home," she said, adding that "the time after Lucy's passing was devastating". She also emphasised the importance of comprehensive, factual, written information being available when the diagnosis is made. "As parents we hear a lot but it doesn't all go in," she pointed out.

SARAH HYNES, of Every Life Counts, conveyed to the audience her "difficult but rewarding" experience following the diagnosis of her son Seán with Trisomy 18 at her 20-week scan in 2015. Sarah told the audience, "I really cannot over-emphasise the importance of good perinatal hospice care for babies like Seán and families like mine."

Seán lived for just two days after birth, but Sarah explained how being able to make memories with her son and take photographs had contributed enormously to the healing process. She also said that the use of the term 'incompatible with life' was misleading and upsetting, and that it made her feel that her son's life was written off before he was born. Sarah told the conference: "Nobody came to help when I asked for assistance with feeding him." When she asked for help, she was told 'try it yourself and if he doesn't take any milk, it doesn't matter'. "His basic human needs weren't being met. Those words will haunt me forever," she said.

"Seán was, and is, such a special little boy. He has had such a huge impact on our lives and we will always remember him in a very special way," she said.

The experiences of each of these parents contributed enormously to the conference, providing a great deal of context for the discussion, and putting faces to the children and families affected by these tragic life-limiting conditions.



Jacinta Murphy
Féileacáin

Sarah Hynes
Every Life Counts

Ide O'Sullivan
Soft Ireland

Dan and Genna Haley
Mam and Dad to Shane

Sarah Nugent
Every Life Counts

Barbara Farlow
International Trisomy

You can watch **parents'** presentations here: www.perinatalconference2016.com

Féileacáin www.feileacain.ie
SOFT Ireland www.softireland.com
Every Life Counts www.everylifecounts.ie
International Trisomy Alliance www.internationaltrisomyalliance.com

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