

The BIT:S (Bronchopulmonary Dysplasia Interdisciplinary Team: Severe) Initiative at Women and Infants Hospital of Rhode Island

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ABSTRACT

Bronchopulmonary dysplasia (BPD) is a major cause of morbidity and mortality in surviving extremely preterm infants, with long-term morbidity disproportionately affecting children with severe BPD (sBPD). Infants with sBPD experience multiple organ system dysfunction. To best treat these complicated patients, we created a multidisciplinary team in 2011 consisting of multiple pediatric subspecialists with a specific interest in sBPD. In the past six years, 150 patients have been referred to our multidisciplinary team, with 131 of the 150 patients discharged home, 65% on home oxygen. Twelve were transferred to the Pediatric Intensive Care Unit (PICU), 3 to a level 2 nursery and 4 died. The multidisciplinary BPD team has standardized the care of children with sBPD and complex medical problems and improved outpatient referral to subspecialists.

KEYWORDS: bronchopulmonary dysplasia, preterm, neonate, chronic lung disease

patients across the US every year.⁵⁻⁹ Within this group of sBPD patients, 4–11% require tracheostomy and mechanical ventilation and 66% are discharged home on supplemental oxygen.⁷ Approximately 2,000 children in the US require home ventilation as a result of sBPD and 8–25% of them have pulmonary hypertension.¹⁰⁻¹² In Rhode Island, the rate of sBPD is about 6% of all children born at <33 weeks.

In 2001, the NICHD defined BPD simply as a chronic pulmonary disease, but there is typically multi-system involvement with many co-morbidities, including neurologic sequelae, pulmonary hypertension, gastrointestinal issues and growth failure. BPD has a multi-factorial etiology and individual susceptibility plays an important role. In the pre-surfactant era, BPD was characterized by airway injury, inflammation and parenchymal fibrosis referred to as old BPD. “New” BPD in the post-surfactant era shows a simplified lung with fewer and larger alveoli due to an arrest of normal alveolarization, with minimal fibrosis, epithelial metaplasia and smooth muscle hypertrophy in the small and large airways accompanied by decreased pulmonary microvascular development.¹³

INTRODUCTION

Bronchopulmonary dysplasia (BPD) is a major cause of morbidity and mortality in surviving extremely preterm infants. The incidence of BPD is anywhere from 25-35% in infants with birthweight < 1500 g.¹ In a large National Institute of Child Health and Human Development (NICHD) Neonatal Research Network study, 68% of infants born at 22-28 weeks gestation who survived developed BPD.² Analysis of a United States (US) nationwide database from 1993-2006 revealed an annual decrease of 4.3% in incidence of BPD with increased length and cost of hospitalization of this population³, but no further improvement has been observed since that time.

The 2001 NICHD definition of severe BPD (sBPD) is the need for $\geq 30\%$ oxygen and/or positive pressure at 36 weeks post-menstrual age (PMA) or discharge, whichever comes first, for infants born before 32 weeks.⁴ The point prevalence of children with sBPD has been estimated to be between 11-58% across different centers around the country. An incidence of 16% for sBPD of all children born <32 weeks estimated nationally amounts to approximately 13,000 new

CREATION OF BIT:S

Due to the multiple systems involved, management of sBPD is best achieved through a multidisciplinary approach. In July 2011, opportunities for improving the care provided to infants with severe BPD at Women and Infants Hospital were identified and the Bronchopulmonary Dysplasia Interdisciplinary Team: Severe (BIT:S) was born. The goals of forming such a team were to: 1) improve overall management of infants with sBPD by taking a collaborative approach, 2) standardize ventilation/oxygenation strategies for infants with sBPD, whose needs are very different from the typical preterm infant, 3) screen for and manage infants with pulmonary arterial hypertension according to published best practice, 4) create a smooth transition from inpatient to outpatient care and 5) establish rapport between subspecialists and the family prior to discharge.

Based on the consensus definition of BPD, criteria for BIT:S referral were defined as any infant who, at 36 weeks PMA, met at least one of the following: 1) evidence of pulmonary arterial hypertension on echocardiogram; 2) fraction of inspired oxygen ≥ 0.3 ; 3) positive pressure ventilation

support in the form of continuous positive airway pressure, nasal intermittent positive pressure ventilation or invasive mechanical ventilation or; 4) requiring high flow nasal cannula flow > 2 liters.

The team consists of a dedicated pediatric pulmonologist, a pediatric cardiologist, a pediatric intensivist, all from Hasbro Children's Hospital, a neonatologist with an interest in BPD, and a pediatric pharmacist from Women and Infants Hospital. Pediatric gastroenterologists are available for consultation in infants with concerns for gastroesophageal reflux, feeding difficulties, poor weight gain, the need for a gastrostomy tube and aero-digestive disorders. Infants who need positive pressure ventilator support at the time of the initial consult, have an absent voice or weak/ hoarse cry, or have stridor/noisy breathing also receive a pediatric ENT evaluation. The BPD team rounds twice a month with the infants' medical team consisting of the on-service neonatologist and neonatology fellow, along with a resident or nurse practitioner, respiratory therapist, nutritionist and bedside nurse. Parents are encouraged to meet the team and be present during rounds. The first patients were seen by the BIT:S team in November 2011.

WHY DO INFANTS WITH SEVERE BPD WARRANT A MULTI-DISCIPLINARY APPROACH?

Infants with sBPD are typically born extremely preterm and suffer from a variety of sequelae of extreme prematurity, of which their chronic lung disease is only one component. Growth restriction and nutritional status affect lung and somatic growth; dysplasia within the lungs leads to impaired vascular growth and pulmonary hypertension; gastroesophageal reflux with recurrent aspiration can worsen pulmonary disease and exacerbate pulmonary hypertension. Prolonged respiratory support often leads to oral aversion and feeding difficulties. The multiple medications these patients receive may interact with and impair many different systems. Neurodevelopmental sequelae are common, in part because developmentally appropriate activities are often precluded by prolonged intubation.

Patients with established sBPD make up a relatively small percentage of any given NICU population, and the strategies employed to manage these problems diverge significantly from the typical NICU patient. Once sBPD and its associated co-morbidities have developed, the management needs to switch from an acute care model aimed at rapid improvement, to a chronic care model with a focus on stability and growth promotion with acceptance of the co-morbidities as the new "normal."¹⁴ This shift is illustrated in the recommendations for ventilator and supplemental oxygen management. No longer is the goal to quickly wean the patient to extubate, or get the patient off oxygen. Instead, the strategy is to ensure adequate respiratory support and

allow for growth, improved interaction with caregivers and development of normal oral motor skills.

Ventilator strategies used in acute respiratory distress syndrome to prevent BPD are very different from the ventilator strategies used in established BPD. The complex, multi-compartmental lung pathophysiology of BPD is different from newborn respiratory distress syndrome and requires much slower ventilator rate, larger tidal volume and much higher end-expiratory pressure.^{15,16} Weaning of support is very slow and deliberate; chronic lung disease does not improve in a matter of days, but rather weeks or months. Children with sBPD need adequate respiratory support in order to grow and to engage in developmentally appropriate activities. Adequate gas exchange is not the sole criterion for adequacy of respiratory support. The infant should be comfortable, without agitation or excessive work of breathing. Too much energy can be expended on respiratory effort, leading to increased energy expenditure, inadequate growth and weight gain which, in turn, impairs pulmonary recovery as well as neurodevelopment.

Meeting the complex needs of this small but challenging population is important as ultimately these children will outgrow most of their problems and eventually leave the hospital, though they often still require some respiratory support at the time of discharge.¹⁷ The transition to outpatient care is an important step, which is facilitated by the fact that the family has an opportunity to establish a relationship with the outpatient providers, such as the BIT:S pulmonologist and cardiologist. In turn, the providers are familiar with the patient, the family and the medical course and thus can provide continuity of care upon discharge.

Because sBPD patients have a high incidence of pulmonary hypertension, a pediatric cardiologist is a key member of the team. They perform screening echocardiograms on every patient at the time of referral and repeat as indicated through discharge and beyond. The presence of pulmonary hypertension is associated with much higher risk of poor outcome and changes how the ventilator and oxygen saturation goals are managed, with higher targeted saturations and more ventilator support recommended.¹⁸ Gastroesophageal reflux and chronic aspiration may worsen lung damage and contribute to pulmonary hypertension. Therefore, a work-up for gastroesophageal reflux is indicated in any infant whose lung disease worsens over time or who is diagnosed with pulmonary hypertension.¹⁹ A pediatric critical care physician is also an integral member of the team, because of expertise in the continued management of infants with sBPD and the fact that some infants become too old to be managed in a newborn ICU and need to transition to the PICU. In addition, because of the high rate of re-hospitalization, there is great benefit in establishing a relationship with the family and aligning management strategies between the NICU and the PICU.

RESULTS

From its inception in November 2011 through December of 2017, 150 infants have been evaluated by BIT:S. Initially the number of referrals increased every year. The last two years have seen the numbers plateau with 27 and 28 patients respectively (Figure 1). The majority of those patients was less than 27 weeks gestation age at birth (Table 1 and Figure 2) and weighed less than 1,000 grams (Figure 3). The patients referred to the team born after 30 weeks gestational age had congenital malformations in the form of genetic syndromes or complex congenital heart disease.

As of July 2017, 131 of the 150 patients were discharged home from the NICU; 12 were transferred to the pediatric intensive care unit at Hasbro Children’s Hospital; 3 were transferred to a level 2 nursery pending their discharge, and 4 patients died prior to discharge from the NICU. One patient died from complications of prematurity after discharge. Sixteen patients required tracheostomies and were discharged home on ventilators and 65% of patients were discharged home on supplemental oxygen (Table 1). Thirty percent (30%) of patients had failure to thrive (FTT) and 60% had clinically significant feeding problems.

Table 1. Population characteristics of patients with severe BPD born from November 2011–December 2017

	N = 150
Gestational age (weeks), SD	26.1 ± 2.4
Birth weight (grams), SD	833 ± 428
Female sex	58 (38.6%)
Race:	
Caucasian	81 (54%)
Hispanic	28 (18.6%)
African American	30 (20%)
Asian	3 (2%)
Not stated	8 (5.3%)
Maternal age (years), SD	28.8 ± 6.4
Tracheostomy at discharge	16 (10.6%)
On oxygen at discharge	96 (65.7%)

DISCUSSION

Over the last two decades there have been great advances in the care of very preterm infants and the rates of survival continue to improve. Despite these advances the incidence of BPD has not changed.²⁰ The focus of neonatology has been on the prevention of BPD, but the literature on how to manage BPD once it has been established is sparse. Creating a multi-disciplinary team focused on the care of infants with fully developed BPD is the first step in studying this disease. We now have six full years of patients enrolled in our program and the beginning of a clinical database that allows us to better understand the problems we are facing.

Figure 1. Number of patients referred to BIT:s by year of birth.

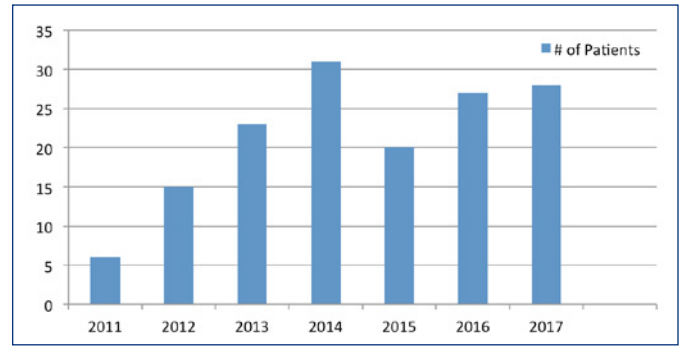


Figure 2. Distribution of severe BPD by gestational age born from November 2011–December 2017

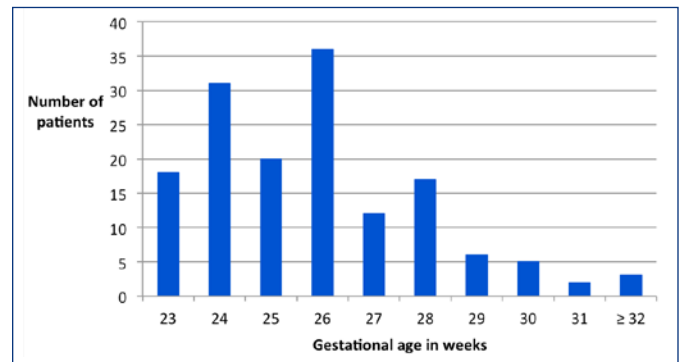
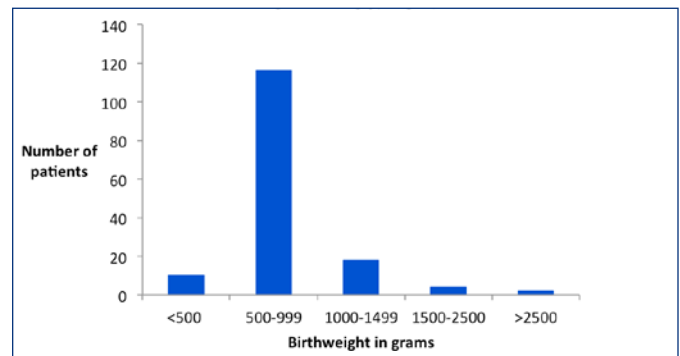


Figure 3. Birth weight distribution in severe BPD born from November 2011–December 2017



We have successfully implemented a multidisciplinary team and now collaborate on the care of every child with sBPD. The number of referrals was initially small, but grew annually between 2011 and 2014 and has now plateaued around 26 patients/yr. As the service has become established, the primary care providers have come to value and appreciate the input of the team as we seek to standardize care and improve best practices. The term “WWBD” (what would BIT:S do?) has even been coined.

One of the goals of the team was to improve the transition from inpatient to outpatient and because all of our patients

are seen by both a pulmonologist and cardiologist while inpatient, there has been an improvement in the transition to outpatient and an increase in the number of follow-up well visits. Nationally there has been a push to create medical homes for complex patient populations and we have been at the forefront of this with our sBPD population. There is a large variability between centers on how sBPD is treated⁷, and because even at large centers the number of patients is relatively small, we were one of the seven founding members of the BPD Collaborative, currently made up of 17 hospitals dedicated to improving the life-long outcomes of babies who develop severe BPD by fostering interdisciplinary collaboration.

With the creation of our own database and of the BPD Collaborative we have put the tools in place to study this complex disease, foster a multi-disciplinary approach and identify areas that have benefited patients and those that need to be improved upon. As an example, when the BIT:S team was founded, gastroenterology (GI) was not a part of it. After our data showed that there were clinically important GI co-morbidities, we have added GI to the team in an effort to improve growth and to facilitate early referral to the Hasbro Children's Hospital's feeding team. Because of the small size of the state of RI and the regionalization of perinatal and pediatric care at Women & Infants Hospital and Hasbro Children's Hospital, we have an opportunity to undertake longitudinal, population-based studies and provide an integrated medical home to this unique and vulnerable population, in an effort to understand the long-term impacts of this disease.

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Disclaimer

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