34 Behavioral Intervention for Positive Airway Pressure (CPAP/BPAP) Desensitization

Keith J. Slifer, Margaret A. Tunney, and Valerie Paasch

Case Vignette

Kenneth was a 3-year-old boy with a genetic condition associated with a range of manifestations including skeletal abnormalities, endocrine abnormalities, developmental delay, and obesity. He was born at 39 weeks' gestation. He had significant respiratory distress shortly after birth and spent several days in the neonatal intensive care unit (NICU) during which time an abnormal heart valve was discovered and he was hospitalized during infancy. Developmentally, Kenneth had limited verbal communication and used only a few simple words consistently. He was ambulatory but had fine motor delays. Moderate obstructive and mild central sleep apneas were revealed on a sleep study at 3 months of age. Continuous positive airway pressure (CPAP) was prescribed but not worn due to Kenneth's inability to tolerate placement of his mask.

Kenneth's bedtime ranged from 8:00 pm to 10:00 pm with 20-min sleep onset latency. He initiated sleep in his own bed but often relocated to his caregiver's bed in the middle of the night. In the morning, he woke around 5:00 am and napped between 4:00 pm and 6:00 pm. Given his inability to tolerate CPAP, Kenneth was referred by his pediatric pulmonologist for CPAP desensitization in an outpatient pediatric psychology clinic at an urban pediatric hospital.

Introduction

Sleep-disordered breathing (SDB), such as obstructive sleep apnea (OSA) [1–5], is more common in children with medical conditions and neurodevelopmental disabilities (NDD). SDB and OSA can lead to disrupted sleep and adversely impact a child's health and daily functioning. Untreated SDB and OSA can impair cardiovascular function and may lead to failure to thrive [1, 6] and difficulties with alertness, attention, and school performance [1, 7]. SDB and OSA that go untreated can also disrupt behavioral and emotional control [7].

Treating OSA in children typically begins with surgery to remove tonsils and adenoids that may be impinging on the child's airway. However, when surgery is not possible or appropriate, or has not adequately treated OSA, positive airway pressure (PAP) is often the next recommended treatment. PAP requires the child to wear an interface, either a mask (attached to headgear consisting of Velcro straps) over the nose and/or mouth or nasal pillows that are securely inserted a little way into the nose. In either case, the interface is connected to the tubing that delivers air at a continuous pressure (CPAP) or bi-level pressure that varies between

K. J. Slifer
Department of Psychiatry and Behavioral Sciences and the Department of Pediatrics, Johns Hopkins University School of Medicine, Baltimore, MD, USA

Department of Behavioral Psychology, Kennedy Krieger Institute, Baltimore, MD, USA

M. A. Tunney
Department of Behavioral Psychology, Kennedy Krieger Institute, Baltimore, MD, USA

V. Paasch (✉)
Department of Behavioral Psychology, Kennedy Krieger Institute, Baltimore, MD, USA

Department of Psychiatry and Behavioral Sciences, Johns Hopkins University School of Medicine, Baltimore, MD, USA
e-mail: paasch@kennedykrieger.org

J. A. Accardo (ed.), *Sleep in Children with Neurodevelopmental Disabilities*, https://doi.org/10.1007/978-3-319-98414-8_34

inhalation and exhalation (BPAP). When successful, this airway pressure is adequate to inflate and keep open the airway to prevent obstructive events. Most PAP machines have a "ramp" feature, set to gradually increase the pressure to therapeutic levels, which is intended to improve tolerance and adherence.

Understandably, PAP adherence can be difficult for children, and their adherence is often suboptimal [8, 9]. PAP requires the child to cope with equipment on his head and face, and even potentially in his nose (nasal pillows), creating unfamiliar and uncomfortable pressure sensations. This is complicated further if the mask is pushed out of alignment as the child changes positions during sleep. Perhaps the most challenging aspect of PAP is that the child must tolerate the sensation of air being delivered under pressure to their nose or mouth, causing additional unfamiliar and initially uncomfortable sensations. The child must learn to acclimate to these sensations, as well as the noise of the PAP machine, and ultimately must be able to initiate and maintain sleep while experiencing these sensations. Prior research has shown that older age and use of a full-face mask decreased CPAP compliance rates [10].

PAP treatment can be challenging for anyone but may be especially difficult for a child with NDD. Children with NDD may have higher rates of disruptive behavior [11], anxiety [12], and tactile defensiveness [13], all of which can complicate their ability to tolerate PAP. They also may have a higher prevalence of facial abnormalities, which can impact proper PAP interface fit. When the fit is poor, the air may blow into the eyes or face, or may shift during sleep, causing discomfort and impacting adherence. While children with NDD are capable of learning to tolerate PAP, they may take a longer time to acclimate to it [10].

Evidence Base for Behavioral Desensitization for PAP Tolerance

Children with NDD may have more difficulty tolerating medical procedures or equipment. Previous research has demonstrated the effective use of behavioral intervention to increase compliance with medical procedures in both typically developing children and those with NDD. Please see the "Behavioral Intervention for Procedural Desensitization" chapter in this book for a more extensive review on the empirical support for procedural preparation in children.

These prior desensitization studies have utilized behavioral-based terminology and interventions, which are briefly defined below:

- *Positive reinforcement*: providing a preferred item or activity (reinforcer) contingent on a desired behavior, such as sitting quietly. Reinforcement is given to increase the chance that the target behavior will be performed in the future. *Differential reinforcement* involves providing a reinforcer in response to behavior one hopes to increase (e.g., sitting quietly), but not in response to behavior one hopes to decrease (e.g., yelling).
- *Negative reinforcement*: removing an aversive or non-preferred item or activity contingent on a desired behavior. This increases the probability that the desired behavior will occur in the future.
- *Escape extinction*: blocking the child's escape from a feared or non-preferred stimulus (e.g., PAP interface) in order to decrease escape behavior that has been maintained by negative reinforcement. Escape extinction is used to teach the child that attempts to remove medical equipment will not be successful; that is, they will not result in escape from the equipment.
- *Counterconditioning*: decreasing negative arousal, such as fear, by using carefully chosen, gradual exposure to the feared stimulus (in this case, medical equipment) while simultaneously engaging the child in a distracting, relaxing, or otherwise pleasurable activity – often a preferred toy or, during daytime rehearsal, a video. The child's anxiety is counterconditioned as he begins to associate the pleasurable activity (toy) with the activity (attaching medical equipment) that previously provoked anxiety.
- *Stimulus fading*: gradually changing some aspect of the physical environment along dimensions of size, shape, color, intensity, proximity to an individual, duration of exposure, etc. while keeping all other environmental variables constant. The goal is to make the change so gradually that it is not noticed by the individual, or if noticed, that it does not change behavior.
- *Shaping*: using differential positive reinforcement to strengthen or "shape" successive approximations of a target behavior. Over time, differential reinforcement of progressively more desirable behaviors, along with withdrawal of reinforcement when unacceptable behavior occurs, teaches the child to behave more appropriately for the situation or procedure, in this case, by cooperating with the steps required for successful PAP usage.

For over a decade, behavioral psychologists have been teaching pediatric patients to cooperate with and adhere to PAP therapy for treatment of OSA and related disorders [10, 14–16]. A 2003 study by Koontz, Slifer, Cataldo, and Marcus [15] involved 20 children with and without NDD who had been diagnosed with OSA and prescribed PAP. Participants either received (1) a 1.5-h behavioral consultation with recommendations; (2) consultation, recommendations, and ongoing behavior therapy (average of three sessions); or (3) consultation, recommendations, and recommendation for ongoing behavior therapy, which was declined by the family. Post-intervention, 75% of those receiving the consultation or the consultation + behavior therapy interventions successfully tolerated PAP and improved their overall usage, as compared to 0% of the group that was offered but declined additional behavioral intervention. Additionally, physicians and caregivers gave high satisfaction ratings regarding the intervention. The results indicated that the children who achieved the greatest increases in mean hours per night of PAP adherence (e.g., >5 h per night) were those reported to have higher levels of estimated cognitive functioning. This initial study was based on a nonexperimental analysis of retrospective clinic data; therefore, a second study was conducted by Slifer and colleagues [16] using repeated-measure, single-participant experimental design.

This second study by Slifer and colleagues used desensitization with four preschoolers aged 3–5 years who had developmental delays and one or more serious health impairments: obesity, heart problems, diabetes, asthma, lung abnormalities, or prior surgery [16]. The children were identified for behavioral intervention because of their distress reactions to the PAP mask and airflow during initial attempts to conduct a PAP titration during polysomnogram (PSG) or to initiate PAP therapy following PSG. Intervention was conducted in inpatient (three participants) or outpatient (one participant) settings and included distraction, counterconditioning, gradual exposure, differential reinforcement, escape extinction, and caregiver training. Behavioral assessment and intervention was conducted using behavioral rehearsal during sessions in which the PAP equipment, mask, and airflow were presented one step at a time.

Before beginning behavioral intervention, each child's mask and equipment were assessed by a nurse or respiratory therapist to determine if the child's mask fit properly. Pressure marks on the skin, air leaks into the eyes, or other discomforts were addressed with interventions including a petite-size gel mask, warm air humidification, use of a "ramp" setting to gradually increase pressure, or other modification to improve the child's physical comfort. Each child was observed during one or two PAP placement attempts. Initial adherence data were recorded and in addition, the child's favorite activities were assessed for potential use to relax, distract, and motivate the child.

A prospective, repeated-measure, nonconcurrent, multiple-baseline experimental design was used to evaluate the behavioral protocol's effectiveness. Prior to behavioral training, none of the children were consistently wearing the PAP equipment. After behavior therapy and caregiver training, all of the children successfully tolerated PAP and increased their hours of use to between 7 and 10 h per night.

Taken in combination, the studies described above provide preliminary empirical support for the usefulness of behavior analysis and therapy in decreasing anxiety, behavioral distress, and escape/avoidance behavior while systematically increasing cooperation and adherence in children with and without NDD having OSA. The combination of task analysis (which will be described in detail later in this chapter), distraction, graduated exposure, counterconditioning, shaping compliance through differential reinforcement, and escape extinction appeared to be effective for increasing child cooperation with and adherence to PAP.

Given the preliminary empirical support for behavioral interventions to improve pediatric PAP adherence, Harford and colleagues developed a systematic program for pediatric patients aged 0–21 with and without NDD [17]. Their program was conducted by specialists and trainees in both clinical psychology and respiratory therapy. Patients were typically seen for their initial appointment following diagnosis of OSA. During this initial appointment, education was provided to the child and caregiver regarding OSA and PAP, a mask fitting was conducted, the child was exposed to and briefly desensitized to the equipment (which continued in additional sessions as needed), and additional strengths and barriers that potentially affect adherence were assessed. The child continued to be seen every 2 weeks for individualized treatment and downloads of PAP adherence data until adherence reached 4 h a night for more than 80% of nights, followed by monthly appointments until adherence was demonstrated for 3 months. Then, patients were reintegrated into the sleep disorders clinic and medically monitored on a regular basis. Preliminary data analysis showed that 5 of the 12 patients following the protocol had greater than 75% PAP usage at their most recent appointment. Barriers to improvement with the PAP protocol included being lost to follow up, depressive symptoms, sensory problems, and lack of caregiver acceptance of PAP. Variable results were found for children with NDD following this protocol. More research is needed on the critical strategies and procedures that are necessary for successful PAP desensitization, as well as on barriers to adherence and creative strategies for overcoming those barriers.

Management

Behavioral training sessions to increase PAP compliance can be conducted on an outpatient or inpatient basis depending on medical status and the urgency for establishing adherence. In either setting, desensitization sessions of 30–60 min with a behavior therapist are typically the most a child can tolerate initially. Parents and other caregivers should be actively involved in the sessions as deemed strategically appropriate by the behavior therapist.

Even though PAP is prescribed for nighttime and naptime use, behavioral training and desensitization *should always occur when the child is awake* in order for the child to receive the full benefit of counterconditioning and to integrate PAP *as an activity that is associated with falling asleep*. This is an important requirement that caregivers often try to skip over. It must be emphasized that children, whose PAP equipment is placed after they fall asleep and have never learned to fall asleep with the equipment on will almost invariably awaken, become distressed by the unfamiliar sensations, and remove the equipment. This can lead the child to avoid going to bed and to become fearful of going to sleep, causing or exacerbating difficulties with sleep initiation and reinitiation after partial nighttime arousals. Attempting to deceive the child by putting the equipment on after falling asleep is almost never successful in the long run.

The first interview with parents focuses on obtaining information about the child's history and previous experience with PAP and other medical diagnostic procedures. In this first session, the child's preferred items and activities should be assessed in order to identify options that may be used for distraction, counterconditioning, and positive reinforcement. The child's own PAP equipment is used during behavioral treatment. This equipment will have been prescribed by the physician and should have been fitted during a pulmonary clinic visit or a visit from a home healthcare provider/durable medical equipment company.

After the initial session, behavioral desensitization sessions follow a task analysis that breaks the PAP into its component steps. Using a task analysis allows for predictable, step-by-step exposure during PAP desensitization trials, in addition to documenting child tolerance and distress behavior during each step. The task analysis may be broken down into written descriptions or picture representation (Fig. 34.1) based on the child's developmental level and preferences.

Sample PAP Task Analysis

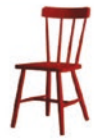
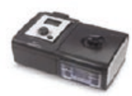
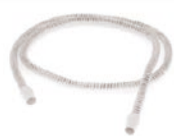
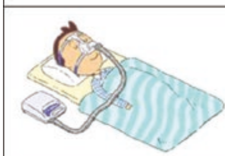
1. Sit on the bed and engage in enjoyable activity.
2. Place mask (not attached to the hose or cap/headgear) on the face for 5 s.
3. Place mask (not attached to the hose or cap/headgear) on the face for 10 s.
4. Place mask (not attached to the hose or cap/headgear) on the face for 1 min.
5. Loosely attach one side of the mask to the cap/headgear.
6. Loosely attach other side of mask to the cap/headgear.
7. On one side of mask, tighten cap/headgear to proper position.
8. On second side of mask, tighten cap/headgear to proper position.
9. Turn on machine/air.
10. Attach tubing to machine.
11. Attach tubing to mask (with pressure turned on) for 3 s.
12. Attach tubing to mask (with pressure turned on) for 5 s.
13. Attach tubing to mask (with pressure turned on) for 10 s.
14. Attach tubing to mask (with pressure turned on) for 1 min.
15. Attach tubing to mask (with pressure turned on) for 5 min.
16. Attach tubing to mask (with pressure turned on) for 10 min.
17. Attach tubing to mask (with pressure turned on) for 15 min.

One advantage of the visual task analysis is that in addition to making the procedure more predictable for the child, it can be used to assist with providing reinforcement (i.e., placing a sticker or stamp once each step is completed, then receiving a prize, or accessing a preferred stimulus or activity after completing all steps included in the session). Additionally, with systematic performance data, the therapist and caregivers can quantify progress that may not be apparent in any individual exposure trial or therapy session. This helps to maintain optimism and motivation to persist with desensitization efforts when progress seems slow.

A key component of this behavioral intervention is counterconditioning sessions to reduce the child's anxiety through gradual exposure to the medical supplies, equipment, and physical sensations the child will experience. This exposure can be conducted while the child is enjoying a distracting, preferred activity, as the activity may keep the child in a relaxed state that overrides or, at least, competes with feelings of anxiety. Once the child visibly relaxes and enjoys the activity, gradual exposure can be conducted by slowly moving the equipment (PAP mask or machine) closer to the child. The duration of contact with the equipment and its sensations (i.e., pressure of the mask or nasal pillow, smell of supplemental oxygen if used and plastic tubing, sound of the PAP machine, air pressure through the mask and into the nose) should be slowly increased, and any cooperative behaviors should be differentially reinforced.

Vicarious learning or modeling can be another helpful strategy. This involves placing the materials on a doll, stuffed animal, or other individual (i.e., caregiver or therapist) and allows the child to become familiarized with the equipment. Caregivers and therapists are able to show the child that equipment is safe, in addition to modeling successful coping, before the child is prompted to wear the equipment. This allows a child to be gradually exposed to the PAP equipment on someone else (and to possibly experience mild distress in doing so), which helps begin to extinguish the child's anticipatory anxiety.

Fig. 34.1 Sample visual task analysis for PAP

	**Sit on the chair**	
	**Practice breathing**	
	**Put mask on nose**	
	**Fasten straps**	
	**Turn on air**	
	**Attach the tube**	
	**Get in bed**	
	**Close eyes**	

Another strategy used in behavioral training for PAP is to allow for gradual exposure to materials (stimulus fading). In addition, in a process referred to as behavioral "shaping," approximations of cooperative behavior are positively reinforced through access to preferred items and activities (i.e., tangible stimuli, videos, games, and social praise). Approximations may begin with reinforcement for sitting in a chair with PAP equipment on a table nearby when prompted, then sitting in a chair with the equipment in the child's lap, then sitting in a chair with one piece of equipment applied, and so forth. In this way, stimulus fading is used for approximating the placement of the mask and the intensity of airflow. The length of time of exposure also is gradually increased as tolerance and cooperation improve.

Escape extinction is implemented as needed by interrupting the child's attempts to pull, remove, or push away the equipment. If equipment is successfully removed by the child, it should be quickly replaced. Blocking escape behavior ensures that the child cannot escape or avoid PAP-related sensations and increases the chances they will begin forming positive associations with PAP resulting from distraction, relaxation, and positive reinforcement (counterconditioning). Additionally, escape extinction teaches children that trials end based on an external cue, such as a timer alerting or a discrete play activity being completed, rather than in response to distress or attempts to remove equipment.

In addition to direct skill implementation with the child, therapists also provide education to caregivers regarding differential reinforcement of cooperation and tolerance of the PAP-related stimuli and sensations while ignoring, redirecting, and blocking escape behavior. Parent training during which caregivers rehearse how to respond to child distress and escape behaviors is important for skill generalization to the home environment.

When the child is able to comfortably tolerate each step of the task analysis, training efforts subsequently focus on increasing the time of exposure to the equipment and sensations. For PAP adherence, the child also must learn to fall asleep wearing the mask or nasal pillow with air pressure at the prescribed level. To promote generalization from the medical setting to the home environment, parents should be assisted with developing a consistent bedtime routine that includes PAP placement. For example, the routine should begin about 30 min before bedtime and should include calming, soothing activities paired with placing the PAP mask and lying down in bed to go to sleep. Direct caregiver training on the intervention procedures should be provided by modeling developmentally appropriate instructions, use of distraction, differential positive reinforcement, and escape extinction as described above. Verbal and written instructions, therapist demonstrations, in vivo behavioral rehearsal with the child, and provision of corrective verbal feedback can be used to train caregivers to generalize these skills to home. When possible, it is helpful to coordinate desensitization appointments with the child's naptime in order to rehearse the bedtime routine and mask placement at a time when the child is likely to fall asleep.

At-home practice may be recommended to assist with generalizing PAP cooperation and coping skills to a different setting. This additional practice can include home-based review of the visual task analysis or practice with PAP materials. This at-home rehearsal allows the child to continue PAP exposure, thereby further decreasing sensitivity and anxiety to the equipment, in addition to giving caregivers the opportunity to identify problems and seek recommendations related to PAP usage at home. As discussed earlier in this chapter, *the ultimate goal is for the child to routinely fall asleep at home with PAP on and air pressure at the prescribed setting. By establishing the association between wearing PAP and falling asleep, caregivers enhance the child's ability to reinitiate sleep with the PAP on during night wakings and partial arousals*.

Case Vignette: Kenneth's Course of Treatment

Kenneth and his grandparents attended an initial evaluation session in an outpatient pediatric psychology clinic at an urban pediatric hospital for children with NDD. In the first session, the therapist obtained behavioral observations and conducted an interview. When presented with a task demand by his grandparent, Kenneth exhibited both disruptive and self-injurious behavior: he whined, flipped a small chair, and hit himself in the face. Undesirable behaviors stopped following removal of task demand and provision of access to preferred items. Information regarding sleep and tolerance of CPAP was obtained. In the 1 year prior to evaluation, grandparents reported that Kenneth tolerated CPAP mask placement for a maximum of 1 min using distraction with preferred activities. Screaming, crying, turning his head, and pushing the mask away were reported during attempted mask placement.

Due to geographic barriers, the family was unable to commute for weekly outpatient sessions. Kenneth was referred for an inpatient admission at a pediatric neurorehabilitation unit for intensive CPAP desensitization. During evaluation, the grandparents reported an average baseline CPAP tolerance of 2 s which could only be achieved with maximum physical assistance. Distress and behavioral dysregulation as evidenced by crying, hitting, kicking, screaming, and hitting himself in the face and head was reported upon presentation of the mask. Similar distress was also reported during use of inhalers and nasal sprays. In addition to CPAP intolerance, general behavioral difficulties noted included disruptive behaviors (e.g., whining, yelling, crying), self-injurious behaviors (e.g., head banging, hitting self in the face and head), and aggression (e.g., kicking, biting, hitting, pinching, throwing objects). Triggers included frustration when denied access to preferred items, soiled diapers, and prompts to complete non-preferred task demands.

Initially, treatment sessions were conducted twice daily for approximately 1 h per session. Kenneth remained seated in his crib during sessions. During the first day of treatment, sessions consisted of 30 trials of 1–5 s duration. His home CPAP equipment was used. The mask was initially disassembled and the pieces were presented separately. Each trial consisted of a task demand paired with a countdown presented both verbally and visually (i.e., fingers on hand, visual timer). Following successful completion of each trial, positive reinforcement in the form of social praise and access to his preferred item, an electronic tablet, was provided. Task demands were systematically increased across trials and included looking at the mask, placing his hand on the mask, and placing the nosepiece on his lower arm, mid-arm, shoulder, top of his forehead, and then nose. Kenneth started to anticipate the steps, demonstrating learning by counting with the therapist using his fingers and pointing to reinforcing stimuli. No crying, physical aggression, or self-injurious behaviors were observed.

On the second day of treatment, trials increased in length up to 15 s with intervention initially remaining consistent. On subsequent trials, positive reinforcement was provided contingent on continued adherence with task demand (i.e., tablet provided during each trial), increasing in length up to 60 s. A visual and verbal countdown was used during the final 5 s of each trial. Task demands were systematically increased to include placement of the mask headgear, first with one strap loosely secured, then two straps loosely secured, and finally all straps secured. Upon achievement of all straps secured, the timer was removed, and trials were continued until Kenneth exhibited verbal distress (e.g., whining) or touched the mask, after which he was prompted to place his hands on his lap and wait for completion of a 5-s countdown. At the end of the countdown, both the mask and access to the preferred item were removed. Once the mask was replaced, access to preferred item was restored. In this way adherence with the mask was consistently paired with access to preferred activities. Kenneth increased tolerance of the mask to 30 min in seven trials. No crying, physical aggression, or self-injurious behaviors were exhibited. He was observed to point his nose toward the therapist and use sign language for “please” to initiate mask placement in an effort to obtain access to his preferred item. For the remainder of sessions throughout admission, when he caught sight of the therapist, Kenneth would sit in the middle of his bed and put a pillow on his lap to prepare for trials in anticipation of the tablet, which was typically placed on his pillow.

During the initial session on the third day of treatment, the therapist collaborated with the respiratory therapist (RT) to evaluate the fit of Kenneth’s mask in relation to the facial anomalies associated with Kenneth’s genetic condition. Due to his history of distress, he never had a proper mask fitting. Upon evaluation by RT, his mask was determined to fit poorly, and a new pediatric mask was provided. Shaping trials increasing in length of time were conducted until tolerance of placement was achieved at which time RT evaluated the fit of the mask. This new mask was also determined to be inadequate so a third pediatric mask was provided. Additional shaping trials were conducted until adequate fit and tolerance were achieved on the sixth day of treatment. No physical aggression or self-injurious behaviors were observed. Two instances of brief distress behavior in the form of crying were observed but resolved with a change to a different preferred activity. Through the shaping trials and differential reinforcement, Kenneth learned that his mask must be kept on in the proper position to gain access to his preferred activity. Because of this, he learned to adjust his mask placement to ensure it was properly placed. When distracted, Kenneth occasionally touched his mask, but in response to a gentle “hands down” verbal prompt, he immediately put his hands on his lap and did not become distressed.

Upon achieving tolerance for mask placement during daytime trials, initiation of mask placement was conducted prior to naptime, and a naptime routine was established (e.g., sit on bed, place mask, provide access to brief video on tablet, prompt to lie down, transition tablet to lullaby music, turn tablet screen away, and maintain audio). Kenneth consistently fell asleep with his mask placed at naptime. After two consistent days of sleep initiation with the mask placed at naptime, it was then also placed prior to overnight sleep initiation. The tablet was faded out and replaced with activities more conducive to sleep: a bedtime story and lullaby music.

Beginning on treatment day 7, daytime trials focused on introduction of air pressure. This began with CPAP pressure set to the lowest possible setting, with approximately 6 in. between the tubing attached to Kenneth’s mask and the tubing attached to his CPAP machine. The distance between the tubing was systematically decreased until tubing was fully connected together. During the initial four trials of full connection, distress behavior decreased with each trial as Kenneth habituated to the CPAP, and no attempts at escape were made. No crying was observed during the remainder of trials. RT remained present during sessions conducted with air pressure and collaborated throughout treatment.

Throughout the remainder of his admission, the therapist provided intervention on approximately 10 additional days. Intervention included daytime trials to increase tolerance of air pressure while systematically increasing CPAP pressures to reach the prescribed pressure target. CPAP mask with prescribed pressure delivered was placed prior to sleep initiation at both naptime and bedtime. The therapist trained caregivers and multiple RTs to complete Kenneth’s naptime and bedtime routine with CPAP placement to generalize to other providers and fade the presence of the behavior therapist. Finally, modifications were made to naptime and bedtime routines to increase efficiency with CPAP placement and decrease breaks between steps. During overnight sleep, some grabbing and pulling behaviors were observed. Blocking was provided to maintain CPAP placement and to promote sleep maintenance and habituation to CPAP. Overnight tolerance of CPAP at time of discharge was approximately 8 h.

Areas of Uncertainty and Future Directions

The procedures detailed in this chapter are based on general principles of behavior and procedures that have broad support in the behavior analysis, behavior therapy, and pediatric psychology literature. Use of simulated (mock) medical procedures, stimulus fading for gradual exposure, distraction, counterconditioning, response shaping using differential positive reinforcement, and escape extinction all have substantial empirical validation in other clinical contexts.

Escape extinction can be initially frustrating to children, but if it is implemented within a multicomponent intervention that includes positive features such as distraction and reinforcement, it tends to be well-tolerated after some initial distress. To be successful, this procedure requires parents and caregivers to allow their child with NDD to experience some mild to moderate distress, as evidenced by crying, pulling away, or shouting. This level of distress is comparable to what a child may experience when frustrated by behavioral demands to participate in non-preferred activities of daily life, such as personal hygiene care, school demands, or bedtime. Children with NDD are routinely exposed to some level of frustration and distress at home, school, and in the community.

More research is needed to test experimentally which of the specific intervention procedures described here are necessary to achieve PAP tolerance and adherence. Controlled single-subject experiments and randomized clinical trials are needed to further refine these interventions. As research accumulates PAP desensitization may be sufficiently manualized for broad application using only essential components, while allowing for individualization for specific individuals with NDD.

Successful PAP desensitization may require the specialized skills of an advanced behavioral therapist working at a specialized pediatric facility. However, with additional training and accumulated practical experience working with individuals with NDD, respiratory therapists, nurses, child life therapists, or other healthcare professionals may be able to successfully desensitize children with NDD to PAP. Ongoing recorded or written data from the training sessions would be helpful. With session-by-session outcome data, behavioral interventions can be individualized to the unique challenges and needs of the specific patient to maximize the chances of success.

Providing the types of behavioral services described in this chapter can be difficult with very young children and with children and youth of any age who have NDD. Also, children with severe anxiety or general behavior problems occurring across settings require an extra degree of behavioral expertise from those who work with them. For children with NDD, the process of therapy can look chaotic at times to the casual observer due to the child's distress behavior and attempts to escape a non-preferred and confusing situation. With more severe NDD, learning may be slower and require more frequent but briefer training trials. Some children may learn more quickly if the training is conducted in brief trials in one setting and then the child is allowed to escape for a contingent break after he or she was cooperative during a training trial. In these situations, the availability of colleagues for backup and "extra hands" may be needed. Specialized environments and resources may be required to keep children safe when escape extinction results in severe tantrums.

Children with NDD may be highly sensitive to routine changes, tactile stimulation, and unexpected noises. However, the behavioral interventions described above can be successful despite these sensitivities but may require more time, resources, planning, problem-solving, and access to the patient and family. Sometimes, when PAP is urgently needed, admission to a pediatric inpatient unit that has intensive medical and behavioral resources available could be considered in places where these resources are available. These types of facilities are most often available at university-affiliated hospitals with special services for individuals with medical, neurodevelopmental, and behavioral disorders. These programs may be referred to as "Behavioral Medicine" programs and may be embedded in departments of pediatrics or psychiatry at major medical institutions.

Finally, professionals attempting to do behavioral desensitization with children and youth with NDD should be conscious of their own capabilities, limitations, and resources before attempting to intervene with individuals with complex neurodevelopmental, behavioral, or psychiatric needs.

PAP Desensitization Summary

1. Children with NDD are more likely than typically developing children to require PAP.
2. Children with NDD are often able to tolerate PAP but may require behavioral training in order to successfully tolerate it.
3. Behavioral intervention can be used to desensitize the child to PAP and to teach cooperation and coping so that distress is minimized and procedural tolerance is acquired.
4. Providers should consider the likelihood that a specific child will be able to successfully tolerate PAP without behavioral intervention, and if tolerance seems unlikely, the child should be referred as soon as possible to a behaviorally trained pediatric psychologist or behavior therapist for desensitization.
5. The initial session should focus on assessing barriers to cooperation (e.g., anxiety, hyperactivity, tactile defensiveness, escape-avoidance behavior) and identify

preferred activities and items to use for distraction and differential reinforcement.

6. The PAP mask and components should initially be introduced during the daytime so distress, tolerance, and progress can be closely monitored while gradually teaching tolerance using behavioral strategies (e.g., distraction, blocking, differential reinforcement) that may be difficult for parents to consistently implement if attempted at home overnight before the child is sufficiently desensitized.
7. The desired outcome is that the child develops a strong association between wearing the PAP and falling asleep so that it will be possible to reinitiate sleep with it on during nighttime partial arousals or night waking.
8. A preferred activity for distraction should be provided during initial introduction of the PAP mask, machine, and airflow pressure.
9. It may be helpful to have the child engage in an activity that is incompatible with removing the mask (e.g., holding a toy or playing a game on the computer tablet).
10. For most children training can be conducted in 30–60 min sessions using their prescribed PAP equipment and materials.
11. Desensitization and training should follow a consistent gradual exposure format by following a task analysis of the behaviors or steps required for successful coping and cooperation.
12. A visual task analysis is helpful to communicate to the child what will happen during PAP and to establish a predictable routine for prompts, exposure, and differential reinforcement.
13. Modeling cooperative behavior can help the child learn the PAP routine and see that it does not cause discomfort or distress.
14. Use stimulus fading to gradually expose the child to the unfamiliar or non-preferred stimuli and sensations involved in PAP while shaping physical proximity and time of exposure.
15. Differentially reinforce successive approximations of cooperation and tolerance of equipment, procedural demands, and sensations using praise and contingent access to preferred items and activities.
16. Gently physically interrupt and prevent escape-avoidance behavior such as pulling off the mask, leaving the area, and hiding one's face while redirecting the child's attention to a preferred distracting activity.
17. Use a timer to communicate the duration of exposure, and signal when the caregiver, not the child, will remove the equipment.
18. Assign time-limited home practice with the specific materials and routine that the child has been able to tolerate in session in order to strengthen learning and transfer treatment outside of the behavioral clinic.

Conclusions and Recommendations

Each child is different and individualization of intervention increases the probability of success. The specific items and activities that will be most effective for distraction and positive reinforcement are idiosyncratic. Selecting the right distractor and reinforcement can dramatically improve the child's motivation to cope, cooperate, and divert attention away from PAP equipment. The behavioral procedures presented in this chapter may not be effective in every case, but with the right distraction and reinforcement during appropriate desensitization and training, the vast majority of youth with NDD can learn to successfully tolerate PAP.

Whenever possible, children with NDD should be referred to a behaviorally trained pediatric psychologist to assess the level of behavioral intervention likely to be required before the child is ready for PAP therapy. Behaviorally trained pediatric psychologists can work with the child, family, and medical caregivers to desensitize the child to PAP and establish environmental modifications and routines necessary to achieve PAP adherence.

References

1. Lal C, White D, Joseph J, van Bakergem K, LaRosa A. Sleep-disordered breathing in Down syndrome. Chest. 2015; 147(2):570–9.
2. Nixon G, Brouillette R. Sleep and breathing in Prader-Willi syndrome. Pediatr Pulmonol. 2002;34:209–17.
3. Jain SV, Horn PS, Simakajornboon N, Glauser T. Obstructive sleep apnea and primary snoring in children with epilepsy. J Child Neurol. 2013;28(1):77–82.
4. Goraya JS, Cruz M, Valencia I, Kaleyias J, Khurana DS, Hardison H, et al. Sleep study abnormalities in children with attention deficit hyperactivity disorder. Pediatr Neurol. 2009;40(1):42–6.
5. Kotagal S, Gibbons VP, Stith JA. Sleep abnormalities in patients with severe cerebral palsy. Dev Med Child Neurol. 1994;36:304–11.
6. Brouillette RT, Fernbach SK, Hunt CE. Obstructive sleep apnea in infants and children. J Pediatr. 1982;100(1):31–40.
7. Beebe DW. Neurobehavioral morbidity associated with disordered breathing during sleep in children: a comprehensive review. Sleep. 2006;29(9):1115–34.
8. Simon SL, Duncan CL. Objective and subjective health parameters and relation to CPAP adherence in pediatric obstructive sleep apnea. Child Health Care. 2012;41:223–32.
9. Marcus CL, Rosen G, Ward SL, Halbower AC, Sterni L, Lutz J, et al. Adherence to and effectiveness of positive airway pressure therapy in children with obstructive sleep apnea. Pediatrics. 2006;117:442–51.
10. O'Donnell AR, Bjornson CL, Bohn SG, Kirk VG. Compliance rates in children using noninvasive continuous positive airway pressure. Sleep. 2006;29(5):651–8.
11. Heffer RW, Chew BL, Perna R, Izmirian SC, Ching MSL, Loughan AR, et al. Intellectual disability and autism spectrum disorder. In: Kapalka GM, editor. Treating disruptive disorders: a guide to psychological, pharmacological, and combined therapies. New York: Routledge/Taylor & Francis Group; 2015. p. 183–206.
12. Grahame V, Rodgers J. Anxiety in neurodevelopmental disorders: phenomenology, assessment, and intervention. In: Van Herwegen J,

Riby D, editors. Neurodevelopmental disorders: research challenges and solutions. New York: Psychology Press; 2015. p. 268–87.
13. Larson KA. The sensory history of developmentally delayed children with and without tactile defensiveness. Am J Occup Ther. 1982;36(9):590–6.
14. Rains JC. Treatment of obstructive sleep apnea in pediatric patients: behavioral intervention for compliance with nasal continuous positive airway pressure. Clin Pediatr. 1995;34(10):535–41.
15. Koontz KL, Slifer KJ, Cataldo MD, Marcus CL. Improving pediatric compliance with positive airway pressure therapy: the impact of behavior intervention. Sleep. 2003;26(8):1–6.
16. Slifer KJ, Hankinson JC, Zettler MA, Frutchey RA, Hendricks MC, Ward CM, et al. Distraction, exposure therapy, counterconditioning, and topical anesthetic for acute pain management during needle sticks in children with intellectual and developmental disabilities. Clin Pediatr. 2011;50(8):688–97.
17. Harford K, Jambhekar S, Com G, Pruss K, Kabour M, Jones K, Ward WL. Behaviorally based adherence program for pediatric patients treated with positive airway pressure. Clin Child Psychol Psychiatry. 2012;18(1):151–63.