



# PEDIATRIC POSITIONING:

## HEY WHEELCHAIR MAN, HOW SMALL CAN YOU GO?

Working in a busy pediatric orthopedic clinic provides daily opportunities for recommending and providing seating and mobility equipment. As the ATP/SMS certified seating specialist<sup>1</sup> at Tampa Shriners Hospital for Children, challenging requests are routinely encountered to meet the orthopedist's goals to correct, prevent or support orthopedic distortions and skin problems, while at the same time ensuring the child's functional ability to propel a manual chair or drive a power chair is not compromised. To ensure favorable outcomes, multiple pieces of information from everyone involved with the child must be considered. What is the optimal outcome? This results when everyone's needs identified in the initial equipment assessment are satisfied. Who is everyone? Not only are the physician and clinician recommendations essential, equally important are the child's needs, the family structure and activities, community activities, transportation requirements, as well as access to the school and home environments. Sometimes, augmentative communication or computer access must also be considered and integrated with the seating and mobility equipment.

Recently, a special child who exemplifies the complexity and multiple needs of the population served in our clinic required a new seating system to meet his functional, positional and orthopedic goals. A 15-month-old male client has a diagnosis of Rhizomelic Chondrodysplasia Punctata Type 1, often called RCPD<sup>2</sup>. RCPD 1 affects fewer than 1 in 100,000 people worldwide and is more common than RCPD 2 and RCPD 3. RCPD results from mutations in one of three genes. Mutations in the PEX7 gene, which is most common, results in RCPD 1. Changes in the GNPAT gene lead to RCPD 2 and, AGPS gene mutations result in RCPD 3. The genes associated with RCPD are involved in the formation and function of structures called peroxisomes.

Peroxisomes are sac-like compartments within cells that contain enzymes needed to break down many different substances, including fatty acids and certain toxic compounds. Peroxisomes are also important for the production of fats (lipids) used in digestion and in the nervous system. RCPD is a condition that impairs the normal development of many parts of the body. The major features of this disorder include skeletal abnormalities, distinctive facial features, intellectual disability and respiratory problems.

Most children with this condition do not achieve developmental milestones such as sitting without support, feeding themselves or speaking in phrases.



PICTURE 1

Convaid Cuddle Bug 2

Affected infants grow much more slowly than other children their age, and many also have seizures.

The child was referred to our clinic by his therapist with concerns about his hip position resulting in decreased range of motion. The family was also concerned about his hands and feet. The child is followed by genetics, receives therapies on a regular basis, and has lately been hospitalized for respiratory issues. He has a gastrostomy tube and has a history of multiple eye surgeries to treat bilateral cataracts.

The medical staff conducted a physical exam of the child and noted shortened humeri and femora, rib cage asymmetry, and bilateral upper and lower extremity joint contractures, particularly at the shoulders, elbows and hips. However, his feet were in a neutral position and were flexible. The child's skin is intact. Due to his preferred hand and wrist position,

**WHAT IS THE OPTIMAL OUTCOME? THIS RESULTS WHEN EVERYONE'S NEEDS IDENTIFIED IN THE INITIAL EQUIPMENT ASSESSMENT ARE SATISFIED.**

upper extremity splints were recommended secondary to right wrist flexion with internal rotation. More significant were the results of the child's X-rays. He had a 30-degree right thoracolumbar positional curve that could be reduced approximately 50 percent with corrective hand forces simulating thoracic lateral supports. The radiograph of the pelvis showed a lack of formation of the femoral heads. The femora were quite widened and shortened with calcifications noted, consistent with his diagnosis. As a result of his clinic visit, the child was referred to occupational therapy for hand splints and for a seating evaluation for improved positioning in his Convaid Cuddle Bug 2 Wheelchair<sup>3</sup>.

Upon initial assessment, a Convaid Cuddle Bug 2 Adaptive Stroller, classified as a compact folding tilt in space wheelchair (HCPCS code of #E1232), would appear to be a good choice for the child with supportive seating (see picture 1). However, the one provided for the child was simply too large to fit his body correctly for upright midline sitting alignment as the seating supports could not be adjusted for his small size (see Table 1).

As a result, the family was forced to keep the seat-to-back angle open and the seating system tilted back to keep the child comfortable and maintain his posture. He was literally looking at the ceiling. A challenge was presented to us to meet the child's positioning and functional needs as this system could not be modified and only a few commercially available products were on the market, which could accommodate his size. One option discussed was a custom sized Jay Sure Fit Lil Kiddos® seating system<sup>4</sup> mounted on a pediatric tilt wheelchair base.

However, this did not meet the family's preference for the stroller base style. We next considered the Otto Bock NUTEC seating system<sup>5</sup>, although when comparing the pricing to our cost to fabricate, we opted to build our own system. The available funding had been exhausted and, with the authorization of the Cuddle Bug 2, those replacement costs would need to be taken care of by Shriners Hospitals for Children. Instead, we decided to offer an in-house custom-made seating system on a commercially available Otto Bock Kimba Spring stroller base that we had in stock.

The custom-made seating insert consisted of a padded I-cut backrest and cushion using an AliMed® T-Foam™<sup>6</sup> one inch soft foam package. Custom-made lateral trunk supports with Lil Kiddos® mount hardware, as well as a custom made anterior trunk support

(CONTINUED ON PAGE 44)



PICTURE 2

The custom seating system interfaced to the Otto Bock Kimba base.

**PEDIATRIC POSITIONING**  
(CONTINUED FROM PAGE 43)

“WITH TEARS IN MY EYES I WANT TO SAY THANK YOU!!!! OUR LITTLE GRANDSON IS 18 MONTHS OLD NOW. THIS IS THE FIRST TIME HE HAS BEEN ABLE TO SIT UP ON HIS OWN (WELL, WITH THE HELP OF HIS NEW CHAIR) AND SEE THE WORLD. YOUR SEAT SPECIALIST AND DOCTORS AT THE TAMPA FACILITY ARE THE MOST AWESOME PEOPLE ... ”

(chest harness) were fabricated. The upholstery covers for the backrest and planar seat cushion consisted of Dartex®<sup>7</sup> on the weight bearing surface. Dartex is a waterproof four-way stretchable breathable fabric that was sewn into Naugahyde side panels and backing to cover the seat and backrest cushions. We installed a Stealth NinÛ head support with a single one-piece sub-occipital pad (QCR #SU118). The head support was attached to the multi-axis removable headrest mount<sup>8</sup> system. We fabricated an adjustable height and depth solid back and seat base with a one-piece footplate (made from Starboard® plastic from the King Plastic Corporation in ¼” thickness<sup>9</sup>. Adjustable width and height lateral trunk supports and adjustable width lateral pelvic supports were incorporated. A custom cutout Lexan® tray<sup>10</sup> was also fabricated (see picture 2).

We estimate eight hours of labor was involved. In addition to the cost of commercial components and fabricated materials, we estimate our cost for the seating insert and tray is \$940. This cost is approximately half of the cost of comparable commercial options



The child three-quarter view.

that were available. The Otto Bock Kimba Spring base was donated from the community for this very purpose.

The custom seating insert was modified on the bottom to accept the Otto Bock quick release system that interfaces to the Otto Bock Kimba Spring base<sup>11</sup>. The Otto Bock commercial canopy was used for those sunny or rainy Florida summer days. Our custom seating system is able to operate exactly the same as the Otto Bock commercial seating system offerings. Pulling the lever on the quick release interface allows us to remove the custom seating system and allow folding of the base. We instructed the family to continue to use the child’s car seat for transportation, as our fabricated seating insert is not crash tested.

Upon initial fit for his new tilt in space seating system, the child was able to sit upright with spinal alignment and maintained midline head, trunk, and pelvic positioning to meet the need for orthopedic support (see pictures 3 and 4). Secondly, because his trunk was in a more extended position his diaphragm had the potential to function more optimally to facilitate deeper breathing and increased air-to-lung volume. This position also benefits digestion<sup>12</sup>. The child now had the opportunity to work on head and trunk control in an upright position and use his hands for grasping and other developmentally appropriate activities with the tray.

The custom chest harness required modification to lower the cross strap and buckle away from his throat. Custom extended plastic tray mounts and elbow pads were made to raise his tray height closer to his elbow height to promote developmental activities. No wheelchair man is perfect.

Feedback ensures growth and learning, which will contribute to improved strategies, techniques and appropriate recommendations of products to meet the needs of our patients. The child’s grandfather wrote to us:

“With tears in my eyes I want to say thank you!!!! Our little grandson is 18 months old now. This is the first time he has been able to sit up on his own (well, with the help of his new chair) and see the world. Your seat specialist and doctors at the Tampa facility are the most awesome people. Y’all have given him something we

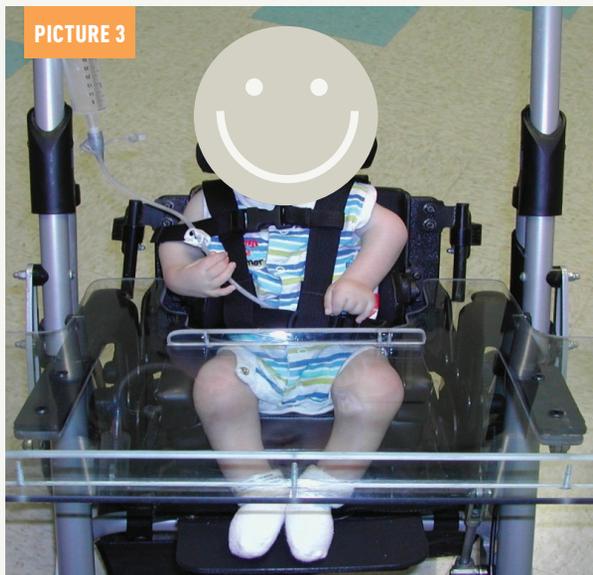
would never have been able to afford. His new chair is just, just ... well I don't have words to say. Again, thank you and all the staff there.”

By using all the information available from each stakeholder and prioritizing everyone's needs, in this case the desired outcome was successfully reached and our costs were ultimately reduced.

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The child on initial fit.

TABLE 1

Measurements		
Dimensions/ Adjustment Range	Cuddle Bug 2 Wheelchair	Rayland's Body Measurements
Seat depth	7-10 inches	4 ½ inch
Seat width	9-12 inches	5 ½ inches
Back Height	17 inches	8 ½ inches
Thoracic pad		3 inches high by 3 inches long
Popliteal to Heel Drop	5-13.5 inches	4 ½ inch
Headrest Adjustment	14-25 inches high	Top of head 14 inches

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