

Evaluation Report of keya

Keya is a sweet 11 year old girl following up for her Physical Therapy rehabilitation for one month. She is a Known Case of SMA Type-2

| Date/Month | Relevant History | Hospital Doctors Visited | Investigations Records | Results and Doctors' recommendation |
|-------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------|
| 22nd December 2010 | Keya was born with her umbilical cord around her neck. FTVD, BCIAB, birth weight 2.25 kg | | No other significant post natal history. | |
| August 2011 (Keya was 8 months old) | Keya's parents noticed that her gross motor skills were regressing when she was 8 months old (reduced limb movements and poor sitting balance). | Neurophysician visited: Dr Shekhar Patil? Physiotherapy started at a different centre. | MRI, EMG, gene mapping, thyroid tests, vitamin D level assessment. | Tests revealed Spinal Muscular Atrophy Type 2. Both parents are carriers of deletion in the SMN1 gene. |
| 30th December 2011 (Keya was 1 years old) | Keya was unable to walk and play with her peers, had frequent respiratory infections | Keya was referred to Dr Snehal Deshpande at Dr LH Hiranandani Hospital by her neurophysician | Muscle charting, neuromuscular and sensory evaluations were performed. Keya was able to come up to sitting, crawl, and pull herself to kneeling. | Keya would require regular physiotherapy sessions that would focus on muscle strengthening and respiratory exercises. |
| 14th march 2012 (1year 3 months) | Keya followed up for therapy 2-3 times per week. She | Re evaluation done at paediatric rehab department at | GMFM- 47.13% Weakness in major lower extremity | AFOs were prescribed and therapy frequency was |

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| | showed improvements in sitting balance, transitions, and could crawl over small obstacles. | Dr LH Hiranandani Hospital | muscles, core and shoulder girdle muscles. | increased to 5 times per week |
| 5th August 2012 | Keya had improved in transitions; she was able to come up to sit, come out of a small bed, but her crawling was reduced | Re evaluation done at paediatric rehab department at Dr LH Hiranandani Hospital | GMFM- 34.8%. Eccentric muscle work and co-activation of postural muscles were poor. | Physiotherapy frequency 5 times per week |
| May 2013 to May 2014 | Keya started physiotherapy sessions at home. | | | |
| May/June 2014 | Keya returned to LH. | Re evaluation done at paediatric rehab department at Dr LH Hiranandani Hospital | -Keya was sitting with propped hands, had poor head holding, increased kyphosis. -pelvic asymmetry -Rolling was the preferred mode of locomotion. -Muscle tone was reduced. -There was asymmetric muscle weakness. -GMFM 19.05% | Bilateral AFOs and adaptive equipment advised-theratogs, fabri foam prowaps. |
| 2014 | Keya relocated to Bangalore with her mother and sister. | She was taking physiotherapy sessions there | | |
| 2019 | Kyphoscoliosis started to develop | Was suggested correction surgery, but could not be performed due to lack of | | |

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| | | funding. | | |
| March 2020-December 2021 | Keya moved with her family to Visakhapatnam. | Keya's mother continued with the home exercise program | | |
| November 2021 | Keya started having acute back pain and difficulty breathing at night. | | | Risdiplam was suggested |
| December 2021- January 2022 | Keya returned to Mumbai for her medical check ups | | | |
| | Follow up | Neurophysician Dr Patil | | Risdiplam 6.6 ml/day lifelong consumption |
| 27 Dec 2021 | For Progressive kyphoscoliosis, Pain, and breathing difficulty | Dr Mihir Bapat | Surgery for scoliosis | CT, MRI, X-ray , PFT , Blood tests |
| 28th Dec | For Risdiplam - Availability and | Dr Anita Hegde | | 2 D echo, PFT, Sleep Study, Bipap , Therapy , chest Pt , Limb Pt , Risdiplam , Psychological Counselling,- Meet Dr Kshitij Chowdhary and Dr Neelu Desai Risdiplam |
| 29 th Dec | | Dr Kshitij Chowdhary | | Requires Surgery, CBC, Total Protein, Urine Routine, PT, aPTT INR, Serum Vit D3, Fitness for Surgery, PFT |

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| | | | | BMD, 2D echo , Review with Dr Aleric |
| | | Dr Neelu Desai | | Same tests |
| 1/1/22 | | Dr Warankar | PFT Allegra, Tanb Montairrle for cold | PFT reports suggested Moderate Restrictive Lung Disease |
| 31/ 12/21 2 D echo | reports done SRCC | | | Normal- No Major deviations, minor due to ribcage shift |
| | | | | Alkaline Phosphatase is on a lower side |
| | | | | VIT D deficiency , Calcium Oxalate crystals in Urine |
| 31/12/21 undisplaced fracture, renal calculus in the lower pole | | Dr Hiren Panwala | CT Spine | CT spine suggested- Severe S shaped scoliosis, dextroscoliosis in the thoracic spine, and levoscoliosis in the lumbar spine, cobbs angle is 62 deg, severe subluxation of the of the Rt hip Joint and retroversion and subluxation of the left hip joint,, Anterior |

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| | | | | Wedging and collapse undisplaced fracture, renal calculus in the lower pole |
| 31/12/21 | BMD spine | | | Use of Bisphosphonate |
| | Breathing difficulty at night | ENT | Enlarged and swollen adenoid glands | Immediate adenoidectomy to increase size of airway passage and enhance lung function |
| | Breathing difficulty at night | Pulmonologist Dr Varunkar | Advised PFT | Diagnosed Restrictive Lung Disease, Keya would require BiPAP support at night |
| | | Endocrinologist | Severe osteoporosis, hypercalciuria | Calcium, vitamin D supplementation with zoledronic acid infusion |
| | | Nephrologist? | Renal Calculi, UTI, bladder retention due to pelvic asymmetry | Surgical removal of calculi |
| | | | Spine, B/L pelvis X ray | Kyphoscoliosis, 50% dislocation of right hip B/L knee flexion contracture Spine correction surgery, implants needed for pelvis and spinal alignment |
| | | Nutritionist | | More alkaline |

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| | | | | diet, stopping animal protein intake |
| 11/1/2022 | | Paediatric physiotherapist, Dr Snehal Deshpande | | Respiratory and limb physiotherapy to improve cardiopulmonary fitness and prevent further progression of deviations in spinal curvature. |

Manual Muscle Testing

| Muscle | LT | RT |
|--------------------|------|------|
| EHL | 3 | 2- |
| EDL | 1++ | 1+ |
| FHL | 1++ | 1++ |
| Dorsiflexors | 2+ | 2 |
| Evertors | 2+ | 2 |
| Tib Anterior | 3 | 3 |
| Knee Flexors | 2- | 2- |
| Knee Extensors | 1++ | 1 |
| Hip Flexors | 1++ | 2- |
| Hip Extensors | 1- | 1- |
| Hip Abductors | ?1 | ?1 |
| Shoulder Flexors | 1++ | 1++ |
| Shoulder Abductors | 2++ | 2++ |
| Trapezius | Fair | fair |
| Elbow Flexors | 2 | 2 |
| Elbow Extensors | 2 | 2 |

| | | |
|-----------------|------|------|
| Wrist Extensors | 3+ | 3+ |
| Intrinsics | poor | poor |
| Finger Flexors | Fair | Fair |

Observations

- Skin is delicate
- Keya has a good will power and is well motivated to do all the exercises
- Back extensors are weak and atrophied
- Reduced chest expansion and excursion

Advice:

- Proprioception for her feet- deep pressure
- Focus on Trunk Rotations and Extensions
- Breathing exercise
- Individual muscle work
- Incentive Spirometer
- Use of Prowraps
- Kinesiotaping
- Good Shoes and Lower Limb orthosis- Static AFO's
- Good booster seat
- Core work
- Working on Hip extensors
- Use of Vibrations for her ribcage with Pillows
- Standing

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