

# LOOK OUT FOR EARLY SIGNS OF SPINAL MUSCULAR ATROPHY (SMA)<sup>1-3</sup>

SMA is a race against time, where fast diagnosis is vital because damage that occurs before treatment is irreversible<sup>4,5</sup>. Check for the signs below at routine check-ups or if parents or caregivers raise any concerns<sup>3,6</sup>.



## SIGNS OF SMA: AGE 0-6 MONTHS

Very early signs of SMA are typically seen up to 6 months of age,<sup>1,2</sup> often by age 3 months.<sup>2,7</sup> Although babies may be symptomatic, they will remain alert and attentive; their cognition not affected<sup>1</sup>



### HYPOTONIA<sup>1,7</sup>

- A baby with hypotonia is often described as 'floppy'<sup>8</sup> due to weakness in their arms and legs<sup>1,7</sup>
- Symmetrical weakness that is more proximal than distal<sup>3</sup> means that a baby will have difficulty lifting their arms and legs, but retain use of their hands and fingers<sup>8</sup>
- The baby's legs may seem weaker than their arms<sup>3</sup>
- In profound cases, the baby may have a frog-like posture when lying<sup>1,9</sup>



### AREFLEXIA<sup>2</sup>

- Absent or reduced deep tendon reflexes are characteristic of SMA<sup>1,2</sup> and a critical part of the baby's exam in cases of hypotonia<sup>9</sup>
- Evaluation of deep tendon reflexes can be achieved by close observation of the baby's response to brisk strikes of the tendon with a specialized hammer<sup>10</sup>



### HEAD LAG<sup>11</sup>

- If a baby seems unable to lift their head or has poor head control,<sup>1,3,8</sup> the pull-to-sit test can be used to confirm head lag<sup>12</sup>
- A baby that is not developing typically will likely have their head lag behind their trunk, with their neck completely extended<sup>11-13</sup>
- They may not lift their head above the line of their back when held horizontally face down<sup>9</sup>



### DIFFICULTY BREATHING<sup>7,8</sup>

- Weakness of the intercostal muscles with sparing of the diaphragm can give the baby a bell-shaped chest and paradoxical pattern of breathing, sometimes referred to as 'belly-breathing'<sup>1</sup>



### DIFFICULTY SWALLOWING<sup>1,3</sup>

- Difficulties with sucking, feeding, or managing oral secretions (saliva) can suggest tongue and swallowing weakness typical in SMA<sup>1,3,11</sup>
- In more progressed cases, a history of choking, recurrent aspiration, or slow or reduced growth may be present<sup>1,8,11</sup>



### TONGUE FASCICULATION<sup>1-3</sup>

- A baby with SMA often will have tongue fasciculations, or twitching of the tongue, together with atrophy<sup>1-3</sup>



### WEAK CRY & COUGH<sup>3</sup>

- A baby with SMA may have a weak cry<sup>3,8</sup>
- Weakness of the respiratory muscles can also cause severe difficulties with coughing<sup>8</sup>

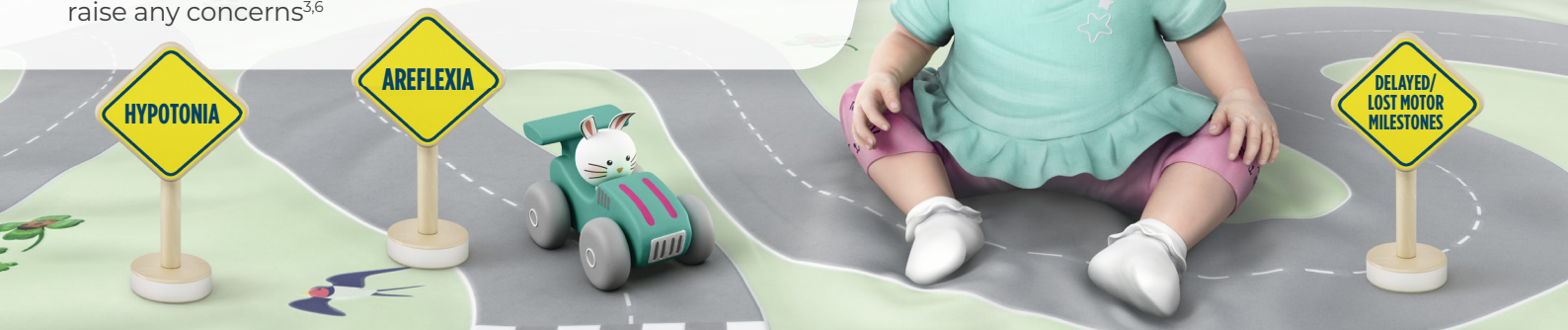
REFER URGENTLY TO YOUR LOCAL PAEDIATRIC NEUROLOGIST IF YOU SEE THE SIGNS<sup>4,14</sup>



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# LOOK OUT FOR EARLY SIGNS OF SPINAL MUSCULAR ATROPHY (SMA)<sup>1-3</sup>

As a healthcare professional you are uniquely placed to spot whether a baby is developing as they should.<sup>6</sup> Check for the signs below at routine check-ups or if parents or caregivers raise any concerns<sup>3,6</sup>



## SIGNS OF SMA: AGE 6-18 MONTHS

Early signs of SMA are typically seen up to 18 months of age,<sup>1</sup> often by age 10 months.<sup>7</sup> Although babies may be symptomatic, they will remain alert with normal speech development<sup>15</sup>



### HYPOTONIA<sup>1</sup>

- Reduced muscle tone and strength on examination, perhaps with a history of poor muscle tone in the first few months of life is a key sign of SMA<sup>2</sup>
- Some weakness in the legs and arms may be present<sup>1</sup>
- The baby may have difficulty reaching for and picking up objects<sup>16</sup>
- The baby is unable to stand due to pronounced leg weakness, and unlikely to walk independently<sup>1,3</sup>



### AREFLEXIA<sup>2</sup>

- Absent or reduced deep tendon reflexes are characteristic of SMA<sup>2</sup> and a critical part of the baby's exam in cases of hypotonia<sup>9</sup>
- Evaluation of deep tendon reflexes, can be achieved by close observation of the baby's response to brisk strikes of the tendon with a specialized hammer<sup>10</sup>



### FINE TREMOR<sup>3</sup>

- When the baby extends their fingers or attempts to grip an object with their hands you may see a fine tremor<sup>3,17</sup>
- Twitching of their shoulder muscles can also present<sup>17</sup>



### PROGRESSIVE SCOLIOSIS & JOINT CONTRACTURES<sup>1-3,18</sup>

- The baby may have more severe motion limitations in the lower extremities than upper<sup>19</sup>
- Progressive scoliosis, most likely C-shaped, contractures, particularly of the knee and ankle, and pelvic obliquity may also be observed<sup>1-3,18,19</sup>



### RESPIRATORY SYMPTOMS<sup>2</sup>

- Restrictive lung disease can result from progressive intercostal muscle weakness<sup>2</sup> particularly if the baby also has scoliosis<sup>1</sup>
- Signs of restrictive lung disease include a reduced total lung capacity and forced vital capacity with preserved expiratory volume<sup>20</sup>



### DELAYED/LOST MOTOR MILESTONES<sup>2,3</sup>

- Although the baby may have achieved milestones,<sup>1-3</sup> it is likely they were delayed.<sup>2</sup>
- Eventually a gradual decline in motor function is observed and some milestones, such as sitting unassisted or standing will be lost<sup>2</sup>

REFER URGENTLY TO YOUR LOCAL PAEDIATRIC NEUROLOGIST IF YOU SEE THE SIGNS<sup>4,14</sup>



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