

## Recommendations for the evaluation of patients with SMA

	Patient visits (days)					
	1	14	28	63	180	Ongoing every 6 months (under SMA-specific treatment every 4 months)
<b>Baseline data</b>	X					
<b>Current medical history and clinical examination*</b> including motor milestones	X	X	X	X	X	X
Case Report Form Nusinersen	X	X	X	X	X	X
Case Report Forms Zolgensma	X		X	X	X	X
Documentation of Risdiplam	X			X	X	X
<b>Physiotherapeutic assessments***:</b> <b>CHOP INTEND</b> <ul style="list-style-type: none"> <li>All children &lt; 2 years of age</li> <li>All patients &gt; 2 years of age without ability to sit</li> </ul> <b>HFMSE</b> <ul style="list-style-type: none"> <li>All patients &gt; 2 years of age with ability to sit</li> <li>If CHOP INTEND score &gt;50: CHOP INTEND and HFMSE****</li> <li>If CHOP INTEND score &gt;60: HFMSE instead of CHOP INTEND</li> </ul> <b>RULM</b> <ul style="list-style-type: none"> <li>All patients &gt; 2 years of age with ability to sit in a wheelchair</li> </ul> <b>6-MWT</b> <ul style="list-style-type: none"> <li>All ambulant patients &gt; 3 years of age</li> </ul>	X			X**	X	X
<b>ALS Functional Rating Scale</b> in adult patients	X				X	X
<b>Pulmonary function*****</b>	X				X	X
<b>Documentation of adverse events</b>	<b>Ongoing</b>					

black= untreated patients; grey = patients under SMA-specific therapy

\* Conducted by neuropaediatrician or neurologist

\*\* Only for therapy with Zolgensma

\*\*\* To be conducted by appropriately trained physiotherapists. Definition of sitting and walking ability according to WHO criteria

\*\*\*\* In patients < 2 years of age, HFMSE is not yet possible due to cooperation. In patients < 2 years of age with CHOP INTEND score > 60, the motor part of the Bayley-III Scales should therefore be performed in addition to CHOP INTEND.

\*\*\*\*\* If the patient is insufficiently cooperative