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Дифференциальная диагностика вялых парезов и параличей верхних конечностей у детей первых месяцев жизни (обзор литературы)

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В статье проанализированы данные литературы, посвященной вялым парезам и параличам верхних конечностей у детей первых месяцев жизни. Указанная патология представляет собой гетерогенную группу заболеваний, имеющих различный этиопатогенез: поражение спинного мозга, плечевого сплетения, периферической нервной системы до уровня плечевого сплетения, а также изолированное повреждение периферического нерва. По срокам возникновения вялые парезы и параличи можно разделить на три группы: антенатальные, интранатальные и постнатальные.

Основным механизмом возникновения данной патологии является интранатальная травма. Более редко вялые парезы и параличи верхних конечностей возникают вследствие антенатальных состояний диспластического и травматического генеза, а также постнатального поражения периферической нервной системы травматического или инфекционного генеза. Врожденные контрактуры верхних конечностей в сочетании с вялыми параличами характерны для ряда генетически детерминированных заболеваний нижнего мотонейрона и врожденных миопатий, внутриутробных повреждений плечевого сплетения и периферических нервов. В статье подробно рассмотрены топическая и дифференциальная диагностика данной патологии, клинические проявления, характерные для каждого периода жизни ребенка, и прогноз заболевания. Данное исследование полезно не только врачам-неврологам, но и специалистам смежных специальностей: врачам-ортопедам, реабилитологам, неонатологам для правильной диагностики патологического состояния, назначения адекватного лечения, а также прогнозирования его результатов.

Ключевые слова: интранатальная травма; вялый парез; паралич; парез Эрба; плечевое сплетение; периферические нервы.

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Differential diagnosis of flaccid palsy of the upper extremities in children first months after birth (Literature review)

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This article analyzes the literature related to flaccid paresis and paralysis of the upper extremities in children during the first months of life. This pathology is a heterogeneous group of diseases with different etiopathogenesis. There are various courses of flaccid paresis and paralysis of the upper extremities in children: damage to the spinal cord, brachial plexus, peripheral nervous system to the level of the brachial plexus, and isolated damage to peripheral nerves. According to the time of occurrence, flaccid paresis and paralysis can be divided into three groups: antenatal, intranatal, and postnatal pathology.

The main mechanism of occurrence of this pathology is intranatal trauma. More rare causes of flaccid paresis and paralysis of the upper extremities are antenatal conditions of dysplastic and traumatic origin, postnatal damage to the peripheral nervous system due to trauma or infection. Congenital contractures of the upper extremities combined with flaccid paralysis are connected with genetically determined diseases of the lower motor neurons and congenital myopathies, intrauterine injuries of the brachial plexus peripheral nerves. This article discusses the issues of topical and differential diagnosis of this pathology, the clinical picture suitable for each period of the child's life, and the prognosis of the disease. This research will be useful not only for neurologists, but also for specialists of related specialties: orthopedists, physiotherapists, and neonatologists for making correct the diagnosis, providing adequate treatment, and predicting its results.

Keywords: intranatal trauma; flaccid palsy; Erb's palsy; brachial plexus; peripheral nerves.

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出生后1个月儿童上肢弛缓性瘫与瘫痪的鉴别诊断（文献复习）

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本文对出生后1个月儿童的弛缓性麻痹和上肢瘫痪的文献资料进行分析。这种病理是一组具有不同病因的异质疾病：脊髓损伤，臂丛神经损伤，周围神经系统损伤到臂丛水平，以及周围神经损伤。根据出现的时间，弛缓性麻痹和瘫痪可分为三组：产前、怀孕期间和产后。

这种病理发生的主要机制是内伤。更罕见的是，上肢的弛缓性麻痹和瘫痪是由于发育不良和创伤性的产前条件，以及创伤性或感染性源性的外周神经系统的产后损害。先天性上肢挛缩合并弛缓性瘫痪是许多遗传性疾病的特征，如下运动神经元和先天性肌病，子宫内臂丛和周围神经损伤。本文详细介绍了这种病理的局部和鉴别诊断，儿童生活各个时期的临床表现特征，以及疾病的预后。这项研究不仅对神经学家有用，也对相关专业的专家有用：骨科医师，康复医师，新生儿医师对病理状况的正确诊断，适当治疗的预约，以及对其结果的预测。

关键词： 内伤；弛缓性瘫痪；瘫痪；欧-杜二氏麻痹；臂神经丛；周围神经。

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论证

出生后1个月儿童的上肢弛缓性麻痹和瘫痪是一组病因病机不同的异质性疾病。其发生可能是由于脊髓、臂丛、外周神经系统损伤至臂丛水平，也可能是孤立的外周神经损伤。

根据出现的时间，弛缓性麻痹和瘫痪可分为三组：产前、怀孕期间和产后。

小儿这种病理的主要原因是臂丛的先天损伤。先天性上肢挛缩合并弛缓性瘫痪是许多遗传性疾病的特征，如下运动神经元和先天性肌病[1]，子宫内臂丛和周围神经损伤[2-4]。继发性的产后上肢周围神经损伤可能是新生儿期机械损伤和感染性疾病的并发症[5-10]。

胎儿损伤通常包括胚胎病变，损害脊髓、臂丛和神经干的结构，以及上肢的关节和肌肉（肌发育不全，先天性水痘综合征，产前臂丛神经和周围神经损伤等）[1，11-17]。

肌发育不全是先天性多发性关节挛缩的一种典型形式，以脊髓前角运动神经元损伤和四肢多发性畸形为特征。临床表现可能类似双侧欧-杜二氏麻痹，伴有先天性僵硬的关节挛缩，肌肉发育不全或发育不全，以及关节血脑带的皮肤收缩[1]。

先天性水痘综合征是胚胎病变，在怀孕期间患水痘的妇女中观察到约2%的新生儿。这种病理的典型症状是皮肤上的锯齿形疤痕，四肢缩短和畸形，对眼睛和大脑的伤害，麻痹症，四肢肌肉发育不良。不同器官和系统损伤的临床症状可以表现为不同程度，有些可能完全不存在。轻瘫与宫内水痘的联系可以基于记忆以及其他表现的存在而假定。病毒学数据可能主要在产后早期阳性[11]。

胎儿期臂丛神经病变的临床表现为弛缓性瘫痪或轻瘫，并伴有先天性上肢发育不全，体积变小，以及关节挛缩。与鼻内臂丛神经损伤的患者相比，患有这种病理的患者，在出生后的头几周，根据神经生理学检查，可以检测到肢体去神经支配的迹象。此外，这些儿童在分娩后早期就发现患肢骨骼脱矿的迹象[12,13]。

在文献中，有一例剖宫产患儿出现弛缓性麻痹和瘫痪，并伴有臂丛损伤，但这种病理的确切发病机制尚未确定[13-15]。宫内臂丛损伤的可能原因之一是由于胎盘血液循环不足而引起的臂丛缺血[16]。羊水收缩也可导致臂丛在子宫内受压，从而形成弛缓性麻痹和上肢瘫痪[17]。

同时观察到桡神经、尺神经和正中神经的损伤，在此病例中，鉴别诊断为臂丛损伤[5]。然而，在出生时就已经有典型的皮肤表现（存在收缩）、四肢萎缩，可以在临床检查时正确地确定诊断。许多作者认为，先天性羊膜狭窄伴桡神经麻痹的预后比单纯神经病变更差[18-21]。文献中描述了由于周围神经受到压迫而导致肌肉无力或完全瘫痪的先天性缩窄病例[20-27]。H. Meyer等人（1941）首次描述了新生儿先天性上肢收缩和手下垂的症状[24]。P.M. Weeks（1982）报告了一例尺骨神经、正中神经和桡神经病变伴先天性上肢狭窄的患者。尽管早期切除了缩窄，但患者没有明显改善。整形手术和神经减压也不能导致长期功能的恢复[20]。N.F. Jones等人（2001）观察了三名患有先天性尺神经收缩和尺神经完全瘫痪症状的患者，经电生理学研究证实。1例患者在孕18周的超声检查中发现这种病理，2例患儿在分娩后立即发现这种病理。尽管早期的手术治疗包括神经整形（两个孩子在3个月大的时候做了手术，一个在6个月大的时候做了手术），术后7个月复查患者，尺神经病变现象持续[23]。

胎儿期上肢弛缓性麻痹和瘫痪的原因是先天性肿瘤和肿瘤样疾病，压迫臂丛或周围神经：先天性血管瘤病，横纹肌样瘤，神经纤维瘤病，颈肌纤维瘤，第一肋骨外生[5，28，29]。与此同时，这些患者没有出生后的外伤史，有些患者在出生后的第一个月就出现了轻瘫，并有一个渐进的病程[5，29]。C. de Turckheim（1991）描述了两例第一肋骨外生并臂丛损伤的病例，损伤定位在臂丛水平[9]。

发生前臂神经压迫缺血性影响的罕见原因包括筋膜室综合征引起的先天性缺血性Volkmann挛缩[30]。这种病理状况的病因与胎儿运动不足和胎儿血液凝血系统的病理有关，但确切的发病机制尚不清楚。新生儿腔室综合征患儿一般出生时皮肤区域水肿、坏死，提示子宫缺血，病理过程开始。新生儿坏疽的特征是肢体主要血管的闭塞，这是鉴别诊断的重点。R. Ragland等人（2005）在16例先天性缺血性Volkmann挛缩症患者中，

表 1 产前出现的四肢弛缓性麻痹和瘫痪的鉴别诊断

病态	临床表现	诊断（方法）
肌发育不全[1]	出生时上肢无力瘫及萎缩，多关节挛缩，双侧性	神经生理（ENMG），体感诱发电位
先天性水痘综合征[11]	出生时上肢出现弛缓性麻痹和萎缩，皮肤上有锯齿状疤痕，四肢缩短和畸形，眼睛和大脑受损	神经生理（ENMG）、体感诱发电位；病毒学检查（在生命的最初几周内）；临床和记忆（母亲怀孕有出水痘的现象）
胎儿臂丛病变[12-17]	出生时上肢弛缓性麻痹和萎缩，关节挛缩	神经生理（ENMG）、体感诱发电位；X线片检查（患侧骨质疏松症）
第一肋骨外生[5，9，29]	出生时或出生后最初几个月出现上肢弛缓性麻痹和萎缩	X线片检查；神经生理（ENMG），体感诱发电位
先天性肿瘤和肿瘤样疾病[5，29]	出生时或出生后最初几个月出现上肢弛缓性麻痹和萎缩	神经生理（ENMG）、体感诱发电位；MRI；US检查
Volkmann氏先天性缺血性挛缩[3，4，30]	出生时前臂皮肤的水肿和坏死。出生时上肢弛缓性远端瘫	神经生理（ENMG）、体感诱发电位；多普勒超声；血管造影
先天性桡神经麻痹[18，31-42]	出生时出现上弛缓性麻痹。肩关节外侧三分之一处特征性皮肤表现：瘀斑、凹陷、皮下结、红斑、皮肤出现硬块诊断	神经生理（ENMG），体感诱发电位
先天性缩窄综合征[5，18-27]	出生时上肢弛缓性麻痹和萎缩，羊水收缩	神经生理（ENMG），体感诱发电位

注：ENMG—神经肌电描记术；MRI—磁共振成像；US—超声检查。

有11例在出生后的长时间内观察到残余的神经损伤[30]。

先天性桡神经麻痹是一种孤立性的桡神经先天性麻痹，非常罕见[18，31-42]。与此同时，臂丛神经损伤的特点是结局的高变异性，孤立性桡神经麻痹，尽管临床表现严重，但完全恢复。由于先天性桡神经麻痹的发生罕见且能自行恢复，其真正的患病率尚不清楚。

X. Song，J.M. Abzug（2015）确诊先天性桡神经麻痹55例[42]。患者的特征是外侧肩中部三分之一的皮肤改变：瘀斑、凹陷、皮下结节、红斑和压实。多数患者患侧肩部中部三分之一处皮肤变色，部分患者皮下结节形成[18，31-39，41]。也有神经功能不经治疗自行完全恢复的病例[19，31]。根据F.S. Alsubhi等人（2011），72%的患者在治疗背景下完全康复通常发生在分娩后的第8周[37]。

胎儿期弛缓性麻痹和上肢瘫痪的鉴别诊断见表1。

鉴别诊断的弛缓性瘫和瘫痪的上肢发展在胎儿期

内伤包括因出生创伤和缺氧导致的脊髓、神经根、神经丛和周围神经的外伤性、缺血性或出血性损伤。

产内脊髓损伤通常是由分娩时脊髓的牵引、过伸和旋转引起的。下颈部和上胸部的病变是臀位表现的特征，而上颈部和中颈部则受胎儿的壁位表现的影响。急性脊髓损伤是由出血（主要是硬膜外）、椎管内损伤和脊髓水肿引起的。罕见的情况是椎骨骨折或脱位以及硬脑膜损伤。弛缓性四分麻痹伴呼吸衰竭和低Apgar评分使怀疑可能是出生后脊髓损伤，这是一个例外诊断。作为额外的检查方法，MRI或CT脊髓造影有助于确定损伤的性质—水肿、出血或缺血[43-45]。

出生后臂丛神经损伤是新生儿弛缓性瘫和上肢瘫痪最常见的原因。这种类型的损伤可以与其他类型的损伤合并：锁骨骨折（10%），肩骨折（10%），颈椎损伤（5%），脊髓损伤（<5%），面部神经、舌下神经和喉返神经麻痹，这决定了疾病的临床特征（10%）[46]。分娩时臂丛神经损伤的发生率为每1000名新生儿中0.4至4例[13，47-49]。70-92%的患者有轻度损伤，可在1-2年的生命中自行恢复[13,50]。臂丛损伤主要有三种临床类型：上Duchenne-Erb型、下Dejerine-Klumpke型和全Kerer型。表2显示了这些类型的临床表现[40]。

臂丛神经病变的其他罕见变异包括：

1) 臂丛单个束或干损伤，患者上肢一个或多个肌肉群无力，这是由于臂丛小群运动纤维损伤所致[5，51]；

表 2 新生儿臂丛神经损伤的临床表现

症状	臂丛神经损伤水平		
	上、中臂丛干，C_5–C_7神经根（轻瘫/欧-杜二氏麻痹）	下臂丛干，C_8-Th_1神经根（轻瘫/Dejerine-Klumpke麻痹）	C_5–Th_1神经根（全瘫/Kerer麻痹）
弛缓性麻痹/上肢肌肉麻痹	近端：三角肌、冈上肌/冈下肌；前臂屈肌（二头肌），旋后肌。C_7—前臂伸肌（三头肌），手指	远端：手部的骨间和蠕虫状肌肉，手指屈肌	近端和远端肌肉
上肢位置	肩内收和内旋	手臂沿着躯干垂下，起来像爪子一样	手臂沿着躯干垂下
活跃的运动	缺乏或不受限制的：肩关节回缩和外旋，前臂旋后和屈曲，可能前臂伸展，手和手指（C_7），但手指保持灵活	缺乏或不受限制的：在手和手指上。肩关节和肘关节灵活	上肢所有关节缺乏或不受限制
被动运动	自由，无痛苦		
肌张力的评估	近端无张力或低血压	远端部位的弛缓或低血压	整个肢体的弛缓或低血压
深层反射	肱二头肌和肱三头肌缺乏或降低	心反射缺乏或降低	缺乏
新生儿反射（先天性）	缺乏或降低巴布金反射，摩洛反射，抓握反射		
灵敏度	在近端区域沿外表面可能降低	在远端区域沿内表面可能降低	敏感性都降低了
伴随的症状	新生儿斜颈，霍纳综合征，膈神经麻痹，患侧营养障碍	霍纳综合征，患侧营养障碍	霍纳综合征，患侧营养障碍

2) 双侧臂丛损伤通常具有不对称的临床表现[5，51–53]。

如果孩子在3个月大后肩部二头肌功能恢复，那么在不丧失肌肉力量或关节活动的情况下，肢体功能很少能完全恢复。产科轻瘫的恢复程度也与病变主题相关：近端轻瘫的预后比全瘫或远端轻瘫的预后好[54]。

孤立性周围神经损伤临床上可表现为臂丛神经束损伤。这类损伤的主要临床诊断标准是没有戈霍纳综合征、无力和由来自这段脊髓的其他神经支配的肌肉失去神经支配，以及来自电生理研究（肌电图、神经肌电图）的数据[5]。为了诊断目的，从孩子出生的第一天就开始进行肌电图、神经肌电图和体感诱发电位。在婴儿早期实施这种治疗的适应症是需要明确脊髓、臂丛和个别周围神经损伤的程度和程度[5，7，8]。

产后出现的四肢弛缓性麻痹和瘫痪的鉴别诊断

孤立性上肢周围神经麻痹，主要是外伤性的，在产后最常见。最常见的桡神经孤立性麻痹，导致单侧手臂活动受限。出生后臂丛损伤引起的桡神经麻痹的特点是臂丛活动活跃，肘关节屈曲，手和手指无伸展时出现握力反射[1，2，4]。尺骨神经的孤立性损伤通常是局部创伤造成的，当手指的内收肌和外展肌力量减弱时，在严重的情况下，患上爪形手畸形。然而，在大多数情况下，在保守治疗的背景下，儿童的神经功能障碍在10天到3个月内得到解决[55]。

对于未分化结缔组织发育不良的患者，在测量血压时由于压迫而不当使用眼压计袖带可能造成桡神经医源性损伤，以及肩部肌肉注射时的神经损伤[40，56，57]。

文献描述了非创伤性单神经病变的孤立病例。S.K. Mahapatra等人（2014）报道了4例脓毒症合并桡神经病变导致肩关节损伤的病例。由于桡神经在解剖学上靠近肩关节囊，当肩关节发炎时，被囊拉伸，神经受压，临床上表现为桡神经神经病。在所有的患者中，手和拇指失去主动伸展，3例患者手的三指骨手指没有伸展。治疗开始后10–21天，病情好转，18–35天，手和手指的伸展完全恢复[58]。

表3列出了产后弛缓性麻痹和上肢瘫痪的鉴别诊断。

表 3 产后弛缓性瘫与上肢瘫痪的鉴别诊断

病态	临床表现	诊断（方法）
围产期缺氧缺血性脑病的中枢肌性低血压综合征[59]	在早期，缺氧缺血性脑损伤可以模拟儿童上肢的弛缓性瘫痪，但其特征是累及上肢和下肢的弥漫性肌性低血压，而肌腱和骨膜反射保留或增加。在严重的病例，弛缓性形式的婴儿脑瘫随后被诊断	NSG，MRI，ENMG
外周肌性低血压综合征[59]	沃德尼格霍夫曼病（1型SMA）：全身性肌肉低血压（张力），肌束，肌腱和骨膜反射缺失，整体运动活动减少	EMG，ENMG，遗传研究
	先天性结构性肌病：全身肌肉低血压，肌腱和骨膜反射缺失，肌肉萎缩，结构性骨骼异常，整体运动活动减少，呼吸障碍	EMG，ENMG，遗传研究，肌肉活检
	先天性肌营养不良：全身肌肉低血压和无力，肌腱和骨膜反射消失，早期肌肉萎缩伴纤维化和脂肪组织肥大	EMG，ENMG，遗传研究，肌肉活检
	遗传性代谢性疾病：全身肌肉低血压，缺乏肌腱和骨膜反射，无力，嗜睡，呼吸障碍，经常抽搐，呕吐，脱水	EMG，ENMG，实验（临床、生化）
假性瘫痪（新生儿发生率为5.9%）[38]	它们是由上肢疼痛或畸形以及臂丛神经的模拟损伤引起的。先天性梅毒的帕罗假性瘫痪：上肢关节被动和主动活动的疼痛和限制与肱骨多处微骨折有关	临床和遗忘（母亲有梅毒病史）；实验（RW）；X线片检查
锁骨骨折[40]	限制肩关节和肘关节的主动运动幅度，水肿，锁骨畸形，触诊时焦虑，肩关节被动活动，手和前臂的功能没有紊乱，上肢没有病理征	X线片检查
肱骨骨折[40]	肩关节和肘关节缺乏主动运动和尖锐哭闹伴被动运动，骨折区组织畸形和肿胀	X线片检查
骨髓炎，上肢关节的关节炎[40]	上肢关节疼痛和活动范围受限，疼痛肢体姿势，局部症状（水肿、充血、高热和软组织酸痛、中毒症状、发烧）	X线片检查；实验室诊断（临床和生化血液测试）；细菌学的诊断
Sprengel病（肩带畸形，肩胛骨高位）[40]	肩胛骨位置的不对称性，肩胛骨的变形，上肢被动外展幅度的限制。肩关节主动活动，但有限（主要是外展）	X线片检查

注：ENMG—神经肌电描记术；MRI—磁共振成像；NSG—神经声像图检查。

结论

当确定出生后几个月的儿童的弛缓性麻痹和上肢瘫痪时，有必要确定这种病理的可能原因，由于这取决于治疗方法，结果和预后的疾病。早期电生理检查使我们能够确定神经损伤的程度和严重程度，并区分出产前损伤与出生后和出生后的损伤。对于不典型的临床表现，包括皮肤损伤、皮下脂肪、肌肉萎缩、四肢发育不全和挛缩、麻痹进展以及没有外伤史的患者，有必要进行神经影像学检查和实验室研究。由于小儿上肢麻痹和瘫痪的预后差异较大，采用多学科的方法来治疗这种病理，包括神经学家、神经外科医生、骨科医生和康复学家在内的专家。

附加信息

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利益冲突。作者声明，没有明显的和潜在的利益冲突相关的发表这篇文章。

作者贡献*O.E. Agranovich*—负责制定研究设计，对文章主题的出版物进行审查，分析材料，撰写稿件文本。*G.A. Ikoyeva, E.L. Gabbasova*—负责对发表的文章的主题进行评论，分析材料，撰写和编辑文本手稿。*E.V. Petrova, V.M. Kenis, AV. Sapogovsky, E.V. Melchenko*—负责对文章主题的出版物进行审查，撰写稿件文本。

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