The startle reflex in children with neuropsychiatric disorders

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Appendix

Culture-specific startle syndromes

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Abstract

The Jumping Frenchmen of Maine is a condition that mainly affected French-Canadian lumberjacks in the North Eastern USA in the 19th and 20th century. The symptoms include excessive startling, sometimes followed by involuntary imitative behaviour and/or obedience to commands. Similarly, Latah (Malaysia-Indonesia) and Myriachit (Siberia) primarily consist of non-habituating hyperstartling but various other behavioural responses like coprolalia, echolalia, echopraxia and immediate obedience to commands may occur. Although no recent studies are available, these culture-specific startle syndromes are considered neuropsychiatric in nature and part of a cultural behavioural pattern.
Definition and history

The Jumping Frenchmen of Maine were first mentioned in an article in the Journal of Nervous and Mental Disease from 1880, written by the American physician, George Beard. Dr. Beard described 50 cases of French Canadians from the Beauce region living in the Moosehead Lake district in Maine. The symptoms and classifications of the nowadays living subjects are very similar to Beard’s description. He described the Jumping Frenchmen of Maine as follows:

“...The individuals were not able to prevent themselves from starting, striking, dropping, jumping and repeating words or sounds once another person startled them with sudden exclamations or commands. Some, when addressed quickly in a language foreign to them, would echo the phrase, even to the point of quoting from the Odyssey or Iliad. If one person was suddenly asked to strike another, he would do so without hesitation, even when it was his mother and he had an axe in his hand.”

“Latah” is originally a Malay term for a broad category of strange behaviour. Literally it means ‘Tiklish’, ‘Jumpy’ or ‘Love-madness. Latah is described to occur in Indonesia and Malaysia but also in certain regions of Africa and Arabia (this separate form differs slightly from the Malay/Indonesian form). It was suggested by Yap that references to Latah in Malay literature date to the 14th century. In 1849, J.R. Logan was the first to write in English about Latah behaviour and described it as un uncontrollable startle reaction. There are only a few recent descriptions. Remarkably, the characteristics of Latah described over 100 years ago and those who were described recently are quite similar. However, the equal sex distribution described by O’Brien in 1884 has changed; Latah is nowadays a predominantly female disorder. The nature of the European presence in the Malayan World during colonial times has probably influenced the occurrence and/or the older reports on the Latah phenomenon. In 1968 Geertz described some Latah women at a party:

“The first latah exclaimed in a loud voice to the second latah, 'Dag!', the Dutch greeting, 'Good-day'. The latah immediately responded, 'Dag!' several times, raising her hand automatically each time. When she paused, the first latah woman started her up again. Then, tired of this game, the first latah cried out, 'Merdeka!', the Indonesian slogan 'Freedom!' and the second latah imitated her, and again repeated it over and over. The first latah then left us to take care of her guests, and the second Latah quieted down. She was a very tense-looking woman, with large nervous eyes, of about sixty. She said nothing unless spoken to. It is the custom in Java at one of these feasts, for all of the guests to urge each other politely to eat, saying over and over again, 'Manga!', 'Please eat!' The second latah had been sitting at the side for some time when she suddenly bursted out —this time without being teased— with 'Manga, manga, manga! Compulsively repeating the polite word and its accompanying
gesture, over and over. People then began to tease her and she grew more rattled, and in this upset condition began mixing obscene words in her speech. At one point she offered a cup of tea to someone, with the words ‘please have some vagina’. The word for tea has something of the same sound as the word for vagina.”

The term Myriachit (sometimes translated also as myryachit, meriachit etc) is derived from the Siberian Tungus word meaning ‘doing something stupid and useless because of sudden fear’. It is also known as ‘Arctic hysteria’ or ‘Olonism’. However, ‘Arctic Hysteria’ originally refers to a broader category of far-northern psychological disturbances. The reports on incidents of Myriachit tend to be folkloric in nature. William Hammond, a surgeon/neurologist, collected stories from American soldiers who had been stationed in Siberia, and he tried to describe the syndrome on the basis of the soldiers’ recollections. Hammond wrote:

“...To annoy him [the patient, a steward of the boat], some of the passengers imitated pigs grunting, or called out absurd names; others clapped their hands and shouted, jumped, or threw their hats on the deck suddenly, and the poor steward, suddenly startled, would echo them all precisely, and sometimes several consecutively”.

Because Hammond did not see the cases himself, the article was severely criticized. Shirogoroff and Jackson also described the syndrome in about the same period, the beginning of the 20th century. Shirokogoroff wrote:

“One day he [the subject] was sitting alone, with his small son, in the wigwam. A knife fell down in front of him (evidently the knife had been in the hanging hunting belt). He seized it and thrust it into his son’s body”.

Clinical features and diagnostic criteria

Clinically, culture-specific startle syndromes consist of a complex of behaviours occurring in a specific cultural group. It includes non-habituating hyperstartling and various other stereotypic behavioural responses like coprolalia, echolalia, echopraxia, “forced obedience” (involuntary, immediate obedience to commands) and hypersuggestibility. As the features of the matching behaviour and forced obedience are prominent it is also referred to as startle-matching syndromes. Compared to the Latah (Indonesia/Malaysia) and Jumping Frenchmen of Maine (US/Canada), the reports concerning Myriachit in Siberia are the least extensive. The clinical details of the three syndromes are given in Table 1. For each society affected, the presentation of characteristics varies only slightly. In addition to these three syndromes there are other rare entities like “Yaun of Burma”, “Bah-Tsche of Thailand”, “Mali-Mali in the
Philippines”, “Lapp panic”, “The raging Cajuns of Louisiana” (related to the Jumpers of Maine”), “Ainu in Japan”, “Leaping ague of Scotland”, “Tigretetier of Abessynia” and “Imanenjana of Madagascar” etc. However, as the literature contains no recent references to them these syndromes are mainly of historical interest. Culture-specific startle syndromes are most often classified as a neuropsychiatric startle syndrome, although some authors claim it is merely a cultural phenomenon.

**Jumping Frenchmen of Maine**

As the responses of the patients were never recorded there are no clear diagnostic criteria other than that the responses are stimulus-induced. The individuals startle excessively to unexpected stimuli and, when startled, may jump, scream or swear, throw objects, strike out at objects or others, assume defensive postures and obey commands (such as ‘dance’, ‘jump’ or ‘run’). Echolalia is also common and echopraxia has been described. There is some disagreement whether coprolalia is part of the syndrome. The swearing following a startle in the Jumping Frenchmen of Maine (also referred to as ‘jumpers’) may somewhat different from the blurtting of obscene language reported to occur in Latah. Some of the behaviours, such as throwing knives, striking hot stoves or jumping into fire, were potentially dangerous or injurious. Jumping is elicited by loud noises, sudden gestures or commands and unexpected physical sensations like the sudden arrival from someone from behind. The subjects are typically adult male lumberjacks from the Beauce region of Quebec or from Maine. In contrast to Latah in Indonesia/Malaysia, the Jumping Frenchmen of Maine usually starts in childhood. In 6 of the 8 cases described by Saint-Hilaire onset coincided with the start of work as a lumberjack. Similar to Latah in Indonesia/Malaysia, anxiety and depressive symptoms are mentioned as comorbidity simultaneously with onset of the disorder. The startle responses become more exaggerated with increased frequency of startling.

**Table 1 Culture-specific startle disorders**

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<thead>
<tr>
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<th>Jumping Frenchmen of Maine</th>
<th>Latah</th>
<th>Myriachit</th>
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<tbody>
<tr>
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<tr>
<td>Excessive startle-like response</td>
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</tr>
<tr>
<td>Coprolalia</td>
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<td>+</td>
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<tr>
<td>Echolalia</td>
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<tr>
<td>Echopraxia</td>
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<tr>
<td>Forced obedience</td>
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</tbody>
</table>
Latah

As in the Jumping Frenchmen of Maine there are no clear diagnostic criteria other than that the responses are stimulus-induced. Clinical features are coprolalia (more specifically, involuntary blurring of sexually obscene words like genitalia), saying idiosyncratically stereotyped things or calling out the name of the thing that excited them (e.g. tiger!) echolalia, echopraxia and compulsive unquestioning obedience when ordered to perform actions which may be ridiculous, improper or even dangerous (“forced obedience”). Usually, the patients are prone to constant teasing by others. Latah is typically evoked by being poked forcefully in the side, loud noises, unexpected or phobic objects (snakes, spiders etc) and possibly merely the presence of Westerners. The individuals are conscious while being Latah and able recollect their behaviour afterwards, but claim to not be able to constrain their actions. Latahs are typically middle-aged women of low social-economical status. However, Latah is also prevalent among homosexual transvestite males. In contrast to the Jumping Frenchmen of Maine, there is an absence of Latah among preadolescent children. Geertz described that the persons severely afflicted with Latah give an appearance of being under extreme tension. Those only mildly afflicted seem no different from the other people around them. The patients described in early recordings are typically servants of European employers but this may be caused by an author’s bias. Onset, often sudden and specific, tends to occur during periods of anxiety, depression, or worry or in a situation in which the person finds himself (herself) wary. Intense life events (e.g. the death of a loved one), being intensely startled by others and a “pre-onset” dream (of a sexual or a frightful nature) have been described as occurring shortly before onset of the symptoms. There may be different types of Latah, including at least a mild form, in which the response is associated with hyperstartling itself, and a more extreme form. One classification consists of the immediate-response Latah (episodes of hyperstartling with or without throwing, dropping, striking out or cursing), attention-capture Latah (episodes in which matching or obedience may occur) and enacted-Latah (a long sequence of absurd-appearing behaviour, even in the absence of adequately startling stimuli). Concerning the latter, several authors wrote that Latah patients have to be separated into genuine cases and those which are basically histrionic and exhibitionistic in nature. Latah has a social function in the Malay/Indonesian society. Virtually anyone in the Malay community participated in the complex of behaviours concerning Latah, either as a Latah, an elicitor of Latah episodes, or as spectator. Simons concluded: ‘to a great extent, it is the behaviour of others towards potential Latah that results in developing Latah, and it is the behaviour of others towards Latah that largely determines the behaviours that Latah perform’. The other way round, Latah patients have a license to mock those about them regardless of relative social status and show conduct considered improper for Malay
women. Lower-status persons are more likely to receive startle-teasing than those of higher status. Although they are often embarrassed, Latahs are not considered either morally or legally responsible for what they do after being startled.

**Myriachit**

Like the Jumping Frechmen of Maine and Latah, there are no clear diagnostic criteria other than that the responses are stimulus-induced. Several clinical features will be discussed. The individual would, when provoked, imitate the actions of others against his will. According to Hammond’s article, Myriachit appeared to involve echolalia and echopraxia only (coprolalia is not reported as part of the syndrome, like it is for Latah patients in Indonesia/Malaysia). However, a famous historical report describes a group of soldiers who repeated both the commands and swearing of a colonel. Shirokogoroff:

‘During a parade of this regiment the soldiers began to repeat the words of command. The colonel grew angry and swore volubly at the men; but the more he swore, the livelier was the chorus of the soldiers repeating his curses after him’.

This is an account of a group of persons becoming infected simultaneously, something which has not been described for Latah or the Jumping Frenchmen of Maine. Reports of group Myriachit led some authors to conclude that Myriachit can be endemic in groups trained to obey, like the soldiers described by Shirokogoroff. Another consequence of mass Myriachit descriptions was that the occurrence of Myriachit was linked to the consumption of a drug. One story describes fourteen soldiers who became liable to fits of echolalia following the consumption of hemp oil given by a sufferer from Myriachit in a settlement in Novokievsky. However, other reports of mass affection do not mention substances. The subjects are mostly female inhabitants of far eastern Siberia, especially Yakutsk. The Koryak, Yukaghir, Chuckchee and the Tungus could all be affected (in other reports Yakut and Samoyed are included), but the latter were more prone as they were less acclimatized to the Siberian circumstances. Similar to Latah, milder and more severe forms were described. The mild form consisted of an obscenity exclaimed after a sudden fright and the more severe form consisted of a hypnotic-like state in which the person would mimic sounds and actions and follow orders. Shirokogoroff:

“*Myriachit is liable to diffusion, fashions and variations, both individual and ethnical and is rooted in the normal psychomental complex*.”

The behaviour was said to be common in areas where the winter was harsh. Further, the pattern is described to become more acute during periods of famine.
Epidemiology
There is no information available other than the condition is very rare. Of the Jumping Frenchmen of Maine less than seventy cases have been described. A decreased prevalence of the Jumping Frenchmen of Maine nowadays compared to earlier times has been attributed to the reduced boredom and isolation experienced by the lumberjacks. According to a study of Chiu published in 1972, the prevalence of Latah is well under the 1 % (of 13,219 Malays, 69 Latah patients were identified). However, the same author estimated that Latah occurred in 15 % of Malay females. In an earlier report of 1924 a number of 300 patients in total were described as seen by Dutch physicians working in Indonesia; 80 % of all the Dutch physicians who were interviewed indicated to have come across a Latah patient. There is no information on the prevalence of Myriachit. It was described to be well known to the Russians. It was estimated that half of the Yukaghir women of thirty or forty and older suffered from a mild or more severe form of Myriachit.

Pathophysiology
Only descriptive, but no functional nor electrophysiological studies have been performed in patients with culture-specific startle syndromes. The neurophysiological background of the excessive startle responses remains therefore, largely unknown. The onset, occurrence and nature of the symptoms in the cultural specific syndromes seem to be influenced by both psychological and cultural factors. There is an ongoing debate on whether culture-specific startle syndromes should be seen as a behavioural phenomenon belonging in the cultural or anthropological realm, or whether it represents a somatic neuropsychiatric disorder in which only the actual expression is open to local cultural influences. There is a striking cross-cultural similarity of behaviours of startle syndromes. The Latah paradox is a term first used by Geertz and refers to the contrast of the connection between Latah behaviour and the norms of Malayo-Indonesian culture on the one side and the occurrence of similar syndromes in completely different cultures on the other side. Therefore, in general an underlying universal neuropsychiatric basis is likely. Yap compared culture-specific startle syndromes to reflex-like fright reactions in German soldiers in the First World War. Heightened arousal or anxiety elicits exaggerated startle responses, and therefore may indeed explain the symptoms. In accordance, anxiety or mood disorder symptoms and negative or intense life events are associated with the onset of culture-specific startle syndromes. However, the behavioural features related to the startle response of patients with culture-specific startle syndromes are clearly more complex than in patients with anxiety disorders. The initial brainstem mediated motor startle response phase, which is roughly uniform from time to time
and from individual to individual, is followed by a secondary phase occurring at a longer latency which shows more variation. This secondary phase contains variable, more complex behaviours, possibly under the influence of psychological factors. It consists of an orienting towards the stimulus source by postural adjustments and autonomic changes and may include emotional and voluntary components. A prolonged series of ‘orienting’ and even vocalizations following a startle response is common and considered normal. Culture-specific startle syndromes may be an abnormal exaggeration of the late, secondary phase of the startle response. The complex, long and non-habituating startle-related behaviour of culture-specific startle syndromes may be considered an ‘orienting exaggeration’ caused by psychological factors. In accordance, a recent observational study described that the startle-associated behaviour such as echolalia and forced obedience could be suppressed. Finally, the behavioural symptoms of the culture-specific startle syndromes may be seen as an exaggerated dependency on the environment for behavioural cues. Such behaviour, in the form of utilization and imitation behaviour, is also observed in frontal lobe syndromes. Frontal dysfunction has the potential for releasing brain-stem centers normally under inhibitory control, e.g. circuits within the brainstem.

Specifically for the French Jumpers of Maine, the occurrence of Jumping Frenchmen is related to specific conditions in lumber camps in the 19th and the beginning of the 20th century. That is, as the isolated life of the French-Canadian lumberjacks was quite boring, jumping seems to have a clear entertaining function. Beard described that after the long day’s work the loggers engaged in mutual tickling punching and startling of the fearful, and he thought that this repeated horseplay eventually resulted in the condition. He felt that the disorder was a ‘fixed psychological state’, ‘a survival of habits’ and ‘a remarkable demonstration of involuntary life’.

Specifically for Latah, Malay/Indonesian cultural beliefs and habits play a role in the etiology of Latah. Geertz mentioned the value for elegant and polite speech, the concern over status, sexual prudery and the dread of being startled as four cultural themes which are possibly related to Latah. It has been suggested by several authors that social repression of the women with often a low social status is an important etiological factor. The Indonesian neurologist Syahrir states that Latah patients demonstrate regression both as a form of protest and to awaken sympathy, acceptance and pity for the person. He argues that rebellion (do prohibited things), dread or fear (for an authorial figure) and conditioning by the environment (attention) may play a part in the development of the symptoms.
Specifically for Myriachit, the symptoms have been attributed to difficulties adjusting to the far-northern environment. Shirogoroff did not regard Myriachit as a disease but as a normal phenomenon. He further states that it has a social function "without the Tungus life would be impoverished". He thought it should not be considered hysteria.

**Prognosis**

The condition is usually chronic. Beard described the Jumping Frenchmen of Maine as lasting throughout life. In Chiu's study of 37 Latah patients, 23 claimed that their symptoms were static, 8 improved and 6 worsened.

**Differential diagnosis and diagnostic work-up tests**

Detailed history taking will exclude Hyperekplexia ('major' form) and other neurological causes of exaggerated startle reflexes (see chapter 2 Startle syndromes). Further, this may reveal the behavioural features typical of culture-specific startle syndromes. EMG studies for the pattern of the startle reflexes in the Jumping Frenchman of Maine have never been performed, so have no diagnostic value.

Gilles de la Tourette saw a resemblance between Latah and Jumping Frenchmen of Maine and the syndrome we know to date as Gilles de la Tourette, and suggested that the syndromes shared a similar etiology. Currently, culture-specific startle syndromes and Gilles de la Tourette are seen as different entities. Tics are suppressible in contrast to the initial startle response in culture-specific startle syndromes. In contrast to externally driven culture-specific startle syndromes the tics in Gilles de la Tourette syndrome is considered due to an internal urge. Hysterical jumps, psychogenic startle and the minor form of hyperekplexia clinically resemble these syndromes, but are not culture-specific. Interestingly, the "minor" form of hyperekplexia occurs in families of individuals affected with the major form of hyperekplexia, an organic condition, and therefore could may also be partly a learned response.

**Management**

In the past both serotinergic and dopaminergic agents have been suggested with little effect. As culture-specific startle syndromes are considered neuropsychiatric startle syndromes, psychiatric treatment is indicated. There are a few brief reports describing the effect of psychiatric treatment on neuropsychiatric startle syndromes. The excessive startle reflexes of one patient described as 'latah' (although he did not
belong to a certain cultural group) was successfully treated by psychiatric therapy including both suggestion (known to be effective in conversion disorders) and exposure elements (known to be effective in anxiety disorders). Similar treatment are described for (other) psychogenic startle syndromes.

Further reading


