The "Other" Primary Headaches

Part 1: The "others" that can cause daily headache

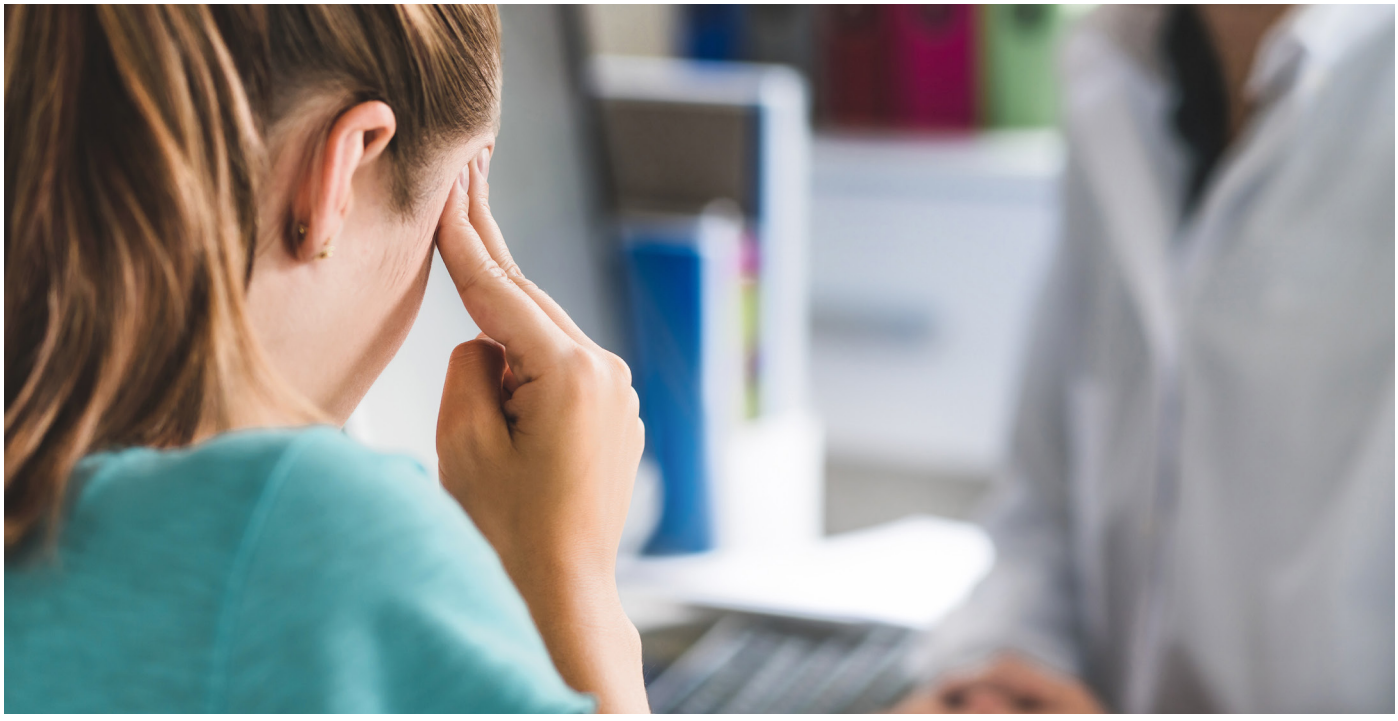
Diagnostically speaking, headache disorders are designated as being either "*primary*" or "*secondary*".

Secondary headaches imply there is an underlying metabolic, structural, inflammatory or infectious process that is causing the headache, and those processes range from something as benign as a self-limited episode of sinusitis to something as grim as an ultimately fatal malignant brain tumor. The first step in headache diagnosis involves ensuring that the patient's headache is not secondary and thus does not reflect disease in the body generally or within the central nervous system specifically.

In the vast majority of cases, however, patients with recurrent headache has a *primary* headache disorder, with *primary* implying that the headache appears to arise in and of itself and not as the result of any underlying disease process. This is a bit of an over-simplification. Strictly speaking, all types of headache disorders, be they *primary* or *secondary*, have a biologic basis and do not simply appear by magic or as some type of psychic phenomenon. For example, while stress may trigger or aggravate migraine, migraine itself is a biologic disorder which generally results from genetic influences not dissimilar in their biological effect from what underlies "primary" epilepsy.

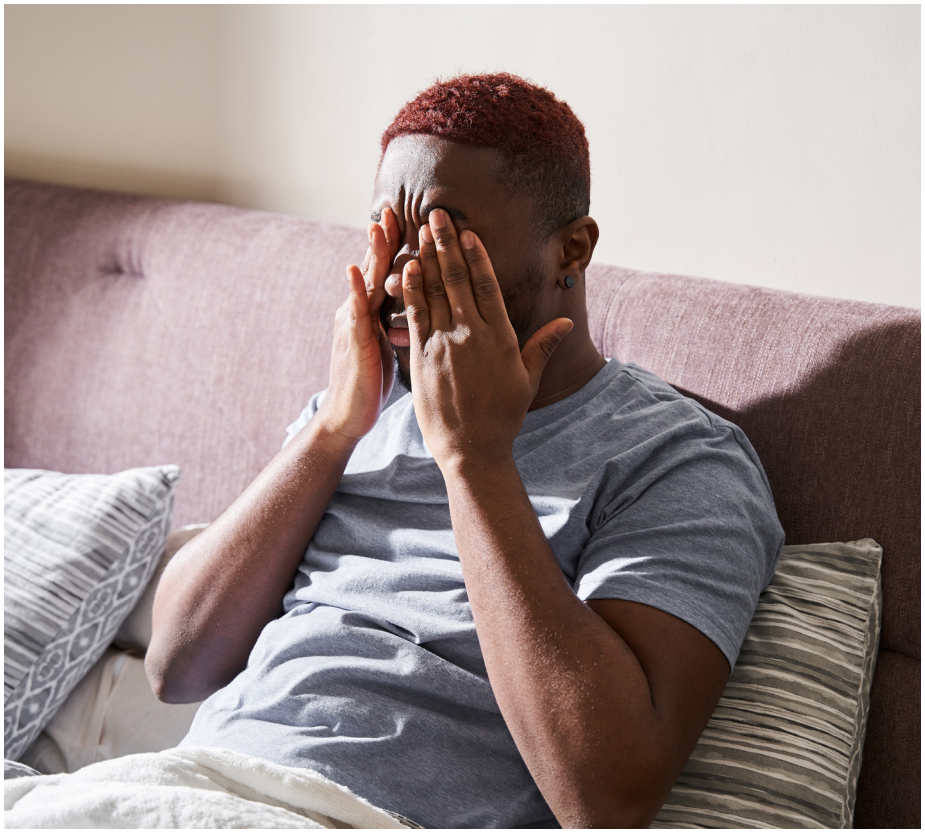
Slightly more than 4% of the American population has headache on a daily or near-daily basis, and the vast majority of those individuals have a primary headache disorder causing their chronic head pain. Although this variant of migraine accounts for only half of chronic headache occurring in the general population, most of those with chronic headache who seek medical attention have chronic migraine.

Most…but not all. No less common than chronic migraine in the general population is ***chronic tension-type headache (CTTH)***, and *CTTH* patients occasionally do show up at the provider's office. Patients with *CTTH* typically do not seek medical attention for their headaches, however, and it is the relative lack of alarming or disabling symptoms that likely accounts for this. In contrast



to migraine, the headaches of *CTTH* tend to be mild to moderate in intensity, rarely incapacitating and free of associated nausea, light/sound sensitivity or aura symptoms. The headache of *CTTH* often is described as "a band-like pressure… like my head is being squeezed in a vise". While migraine's genetic predisposition and biologic circuitry have been fairly well delineated, the same cannot be said for CTTH. Although there must exist some biologic basis for CTTH, our inability to identify this headache disorder's biologic underpinnings not surprisingly has resulted in an absence of evidence-based therapies for its management.

Along with the occasional patient with CTTH, healthcare providers may see patients with less common primary headache disorders that also can cause daily head pain. Ranking first amongst these "others" is ***cluster headache***, much less common than chronic migraine or CTTH but more common than the other primary headache disorders subsequently to be described.



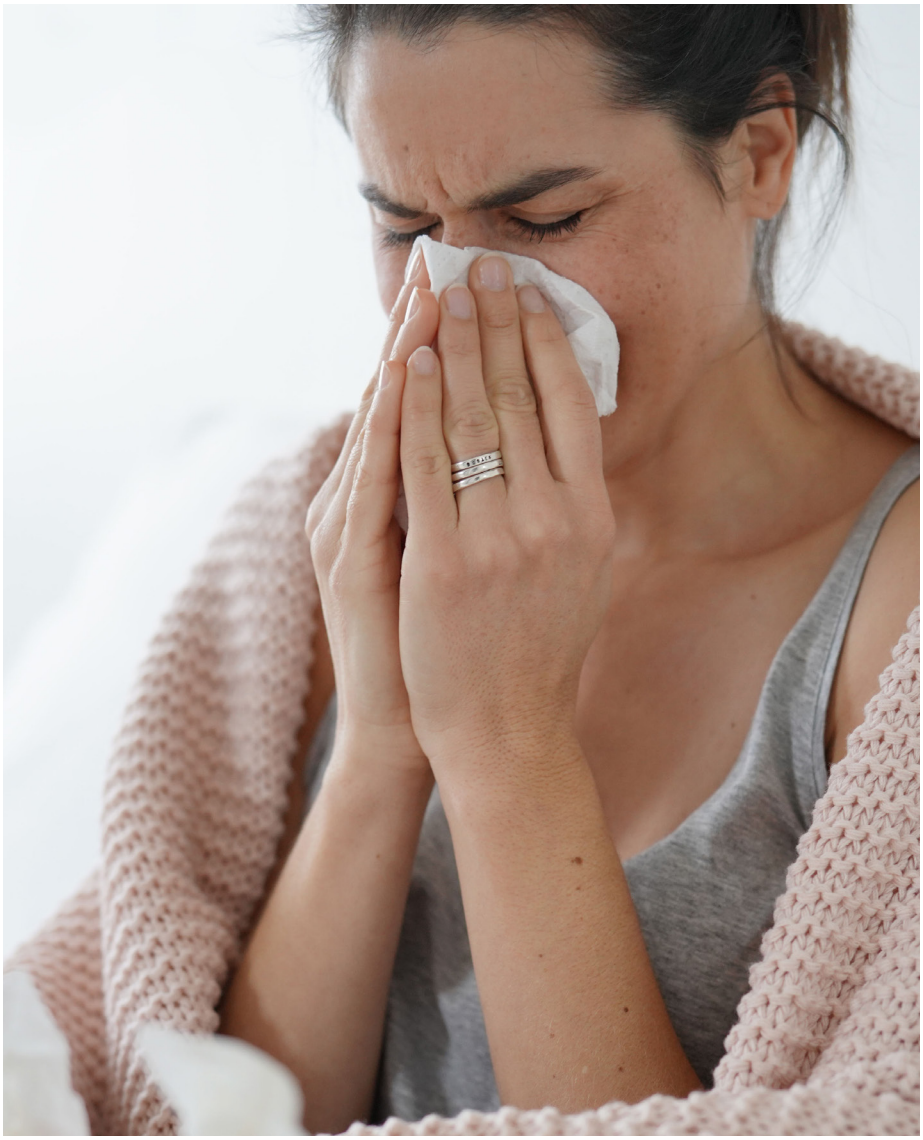
Cluster is a symptomatically vivid headache disorder involving multiple bursts of excruciatingly severe headache that thankfully last less than 3 hours and often less than an hour. Cluster most often is cyclical, with weeks to a few months of up to 6 attacks per day alternating with attack-free intervals that may last for years. When cluster activates, the attacks of head pain are typically "side-locked", right or left, from attack to attack and even cycle to cycle. Acute attacks are often accompanied by tearing, nasal congestion and eye redness on the same side as the headache. Although cluster at times may be misdiagnosed as "sinus headache", a dental problem or something else equally far off the mark, the severity of the pain and the stereotyped nature of the symptoms usually combine with the patient's understandably determined search for relief to result eventually in an accurate diagnosis.

> **Patients with chronic tension-type headache typically do not seek medical attention.**

Paroxysmal hemicrania is similar to cluster in its symptomatology. Like cluster, it tends to occur in cycles of daily headache, is typically side-locked to the right or left, involves headache attacks that are severe in intensity but short in duration, and is characterized by tearing/nasal congestion/red eye on the same side as the headache. *Paroxysmal hemicrania* differs from cluster in that it is about 50 times less common than cluster, females are just as likely to be afflicted as males (cluster occurs much more often in males), there tend to be more attacks per day (more than 5) and the attacks tend to be of shorter duration (2-30 minutes). Perhaps most critical to making an accurate diagnosis of paroxysmal hemicrania is the disorder's absolute responsiveness to indomethacin, a venerable nonsteroidal anti-inflammatory drug that was first approved for clinical use in the US in 1965. When an adequate dose of indomethacin is taken 3 times daily by a patient with *paroxysmal hemicrania*, the therapeutic response is dramatic in its rapidity and thoroughness. This does not occur in cluster patients treated with indomethacin, and unfortunately other NSAIDs which may be better tolerated than indomethacin by a patient with *paroxysmal hemicrania* do not produce the same positive result.

Hemicrania continua (HC) migraine is easiest to think of as "paroxysmal

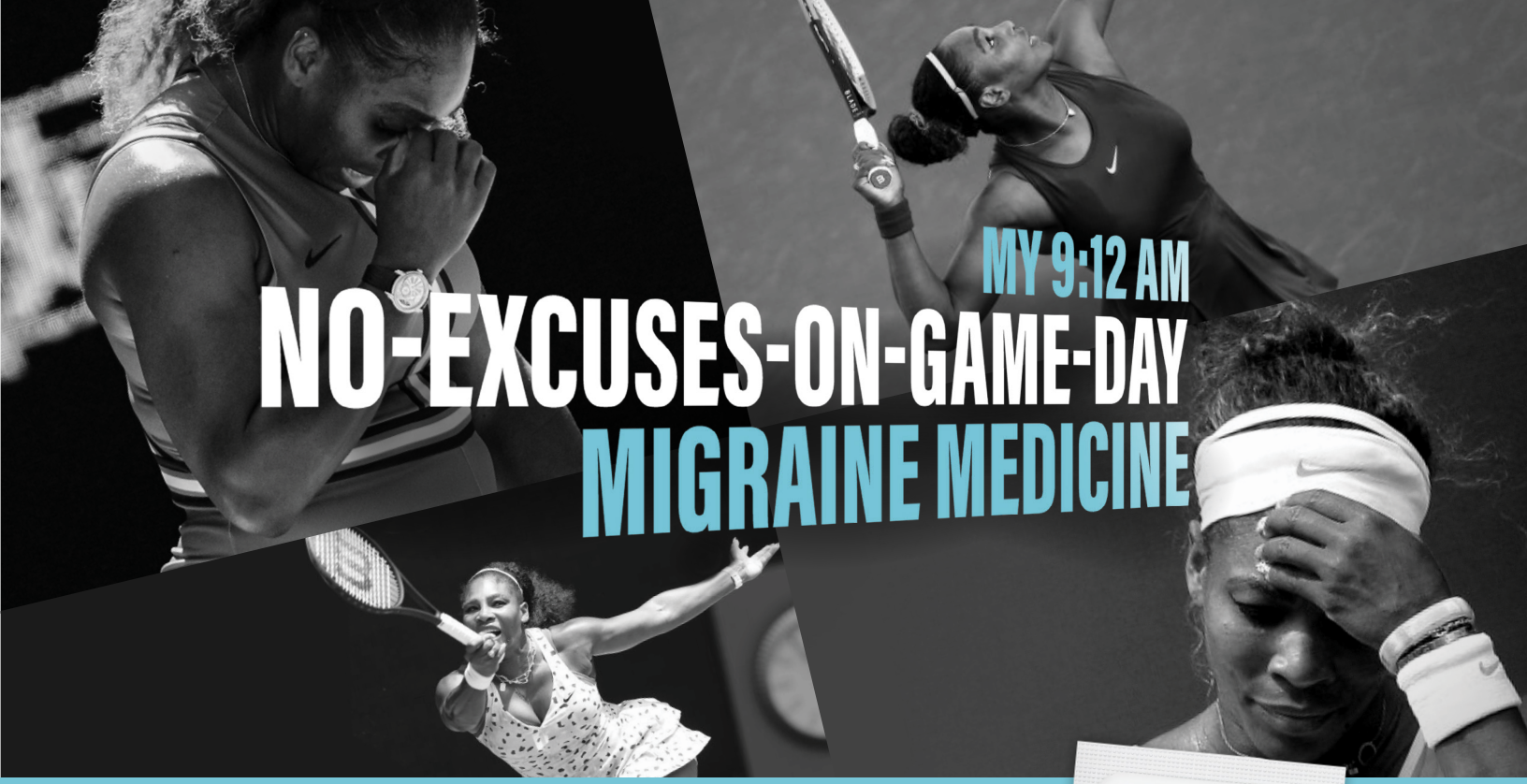
hemicrania that never stops". Individuals with *HC* describe constant "background" head pain that is side-locked to right or left and at times intensifies, with such intensifications lasting anywhere from minutes up to several days. Similar to both cluster headache and paroxysmal hemicrania, during these intensifications of *HC* the individual may experience tearing, nasal congestion, eyelid droop or swelling and red eye on the same side as the headache.* Some individuals with *HC* report having the sensation that there is a foreign body in their eye on the same side as the headache. As with paroxysmal hemicrania, the response to treatment with indomethacin is both absolute and dramatic. Finally, because variants of HC do occur (egs, head pain lateralized to one side but shifting from one side to the other; occasional periods of freedom from head pain; occasional absence of autonomic* features), many headache subspecialists have a low threshold for beginning patients with persistent lateralized headache on a short trial course of indomethacin.



Because such symptoms as tearing, nasal congestion or a runny nose may be generated by the autonomic (involuntary) portion of the nervous system, and because the trigeminal nerve is implicated in producing the head pain itself, *HC*, *cluster* and *paroxysmal hemicrania* are considered to be part of a group of primary headache disorders referred to as the ***trigeminal autonomic cephalalgias*** (TACs). Identification of a patient's headache disorder as being a TAC will avoid both the harm that so often results from any inaccurate diagnosis and, especially in the case of *HC* or *paroxysmal hemicranias*, the failure to treat appropriately a highly treatable headache disorder.

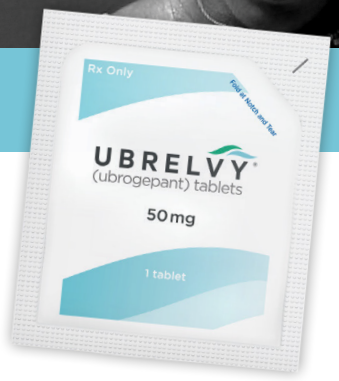
Not a TAC and perhaps not even best classified as a primary headache disorder is so-called ***new daily persistent headache*** (NDPH). Patients with *NDPH* typically have no pre-existing history of recurrent headaches and report experiencing they often abrupt onset of an uncharacteristic headache that persists and never entirely goes away. Often the patient can recall the date - even the precise moment - and under what circumstances the headache began. The "clinical phenotype" (the symptoms and features of the headache) usually mimics either CTTH or chronic migraine (in which case it may be characterized as *NDPH* "with migrainous features"). While this same headache pattern may occur following head injury, thrombosis (clotting) of a large vein within the brain, leakage of cerebrospinal fluid, stroke or central nervous system infection (meningitis, encephalitis), the hallmark of primary *NDPH* is the inability to identify any obvious reason for this chronic headache to have begun in the first place. Headache subspecialists often treat *NDPH* in the same manner and with the same medications the use for patients with chronic migraine. Unfortunately, NDPH is notoriously difficult to treat effectively, and combining this with the inability of even extensive diagnostic testing to identify any underlying "cause" can make *NDPH* a particularly frustrating disorder for patient and healthcare provider alike.

Chronic tension type headache, cluster headache, paroxysmal hemicrania and new daily persistent headache. These are the "other" primary headache disorders which, like migraine, can cause the afflicted individual to experience headache on a daily and even constant basis. In the next issue of this magazine we will discuss the "other" primary headache disorders that are much less likely to occur daily and more likely to be "hit and miss".



MY 9:12 AM NO-EXCUSES-ON-GAME-DAY MIGRAINE MEDICINE

UBRELVY CAN QUICKLY STOP MIGRAINE IN ITS TRACKS.



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—Serena Williams

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Who should not take UBRELVY (ubrogepant)?
Do not take UBRELVY if you are taking medicines known as strong CYP3A4 inhibitors, such as ketoconazole, clarithromycin, itraconazole.

What should I tell my healthcare provider before taking UBRELVY?
Tell your healthcare provider about all your medical conditions, including if you:
- Have liver problems
- Have kidney problems
- Are pregnant or plan to become pregnant
- Are breastfeeding or plan to breastfeed

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