Update Article

Optimising Primary Headache Management

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Abstract

Despite the fact that headache is one of the commonest of medical complaints seen in practice, most headache patients continue to be suboptimally managed. Probably because recent advances in the field of headache are written about more in specialty journals, many physicians even today feel that nothing much can be done for their headache patients! And, since more than 90% of headaches seen in practice are primary headaches, we need to realize that there is enough evidence now to show that primary headaches are a potentially treatable biological problem. It is therefore important that we do not err in the diagnosis and choice of the right drug options. This article discusses some of the reasons for suboptimal management of primary headaches and suggests measures to avoid these 'Pitfalls'. Secondary headaches where there is an underlying cause identifiable on examination or investigation are outside the scope of this review. ©

Introduction

The International Headache Society Classification of Headache Disorders (ICHD-2)[1] divides all headaches into two broad categories. **Primary headaches** are those headaches where examination and investigation are normal and **secondary headaches** are those where the underlying cause of the headache is identifiable on examination or investigation. Regardless of the fact that the pain can be just as severe in both, we are better trained to diagnose and treat secondary headaches and when it comes to dealing with the more common and chronic primary headaches like migraine or cluster headache, most of us falter in the same fashion.

Why this state of affairs? Is it because we have not been trained adequately to recognize these head pain disorders and manage them effectively? Is it because most of these headaches are written about in rare specialty journals? Or is it because these head pain disorders are not as 'interesting' and do not show up with a lesion on the scan or a deficit in the patient? Based on lessons learnt from dealing with difficult headache patients who attended a specialty Headache Clinic in a tertiary care hospital, this review aims at providing a comprehensive check-list for some of the overlooked aspects, hurdles in the management of primary headaches. Secondary headaches where the diagnosis is obvious on examination or investigation will not be discussed here. These 'Pitfalls' which lead to sub-optimal management are listed below.

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1. The true misery of headache is not always understood in a busy out-patient practice.

Headache is an invisible misery, where quite often nothing is seen from outside and nothing is measurable on the inside. The size of the problem, the extent of the misery, the level of the disability and the true burden of headache is generally not known. Most of us concentrate on patients with deficits on examination or lesions on imaging. Almost 95% of headaches seen in practice are primary headaches but still they are not considered a serious problem warranting great attention.

Because they are never life-threatening, because they do not cause permanent disability and because there is no focused teaching on 'primary headaches' in the undergraduate and postgraduate curriculum, primary headaches continue to stay neglected without specific diagnosis and treatment. As a result, there is a huge gap between the theory and practice of headache medicine leading to suboptimal management and needless frustration for both patient and physician.

2. Headache is a common complaint, the cause of which overlaps many specialties.

With so many different causes for headache and with an overlapping nerve supply to the head and face, it is not surprising that headache patients are shunted between various specialists, who look at the problem through the window of their own specialty. Mismanagement of headache takes place at various levels. Many headache patients make the mistake of self-medicating because of fixed ideas, myths, and the mistaken notion that nothing more can be done for their headaches. Because of overcrowding, family physicians are not in a position to take a detailed headache history. Internists are more

Table 1 : The International classification of headache disorders

Primary Headaches Disorders

1. Migraine
   - 1.1 Migraine without aura
   - 1.2 Migraine with aura (6 subtypes)
   - 1.3 Childhood periodic syndromes that are commonly precursors of migraine (3 subtypes)
   - 1.4 Retinal migraine
   - 1.5 Complications of migraine (5 subtypes)
   - 1.6 Probable migraine (3 subtypes)
2. Tension-type headache
3. Cluster headache and other trigeminal autonomic cephalalgias
4. Other cluster headache

Secondary Headache Disorders

5. Headache attributed to head and neck trauma (7 subtypes)
6. Headache attributed to cranial or cervical vascular disorder (7 subtypes)
7. Headache attributed to non-vascular intracranial disorder (9 subtypes)
8. Headache attributed to substance or its withdrawal* (4 subtypes)
9. Headache attributed to infection (4 subtypes)
10. Headache attributed disturbance of homeostasis (7 subtypes)
11. Headache or facial pain attribute to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth mouth or other facial or cranial structures (8 subtypes)
12. Headache attribute to psychiatrist disorder (2 subtypes)

Headache Classification Committee of The International Headache Society. The International Classification of Headache Disorders (second edition). Cephalalgia2004; 24: 1-160

used to diagnosing and treating the typical presentations of migraine and tension-type headache, but not complex headaches. Overlap specialists tend to overdiagnose in their own specialty - the ENT surgeon labels it sinusitis associated headache, the ophthalmologist attributes it to the minor error in refraction, the gastroenterologist who is consulted for the dyspeptic symptoms treats only what is seen on endoscopy, the gynaecologist wrongly labels menstrual migraine as pre-menstrual syndrome, the orthopaedic surgeon who is consulted for the neck pain of migraine calls it cervical spondylitis and the psychiatrist is eventually the last halt or the final bouncing board! Unfortunately as a study from our clinic revealed , the neurologist who is the specialist most suited to handle the difficult headache patient, is consulted by less than 10 % of headache patients. In India, the ophthalmologist still remains the consultant of first choice for most headache patients.[2]

3. Secondary headaches comprise less than 10% of headaches seen in practice!

It is well known that more than 90% of headaches seen in practice are primary headaches and in these patients there is nothing abnormal found on examination or investigation. Primary headaches are more common, more chronic and also more difficult to treat. The severity of the pain may be just the same regardless of whether or not there is an identifiable underlying problem. But probably because secondary headaches show some abnormality, have a specific treatment, and are easier to manage, most clinicians are adept at diagnosing secondary headaches, but are uncomfortable dealing with primary headaches. This is compounded by the fact that most textbooks outline clear guidelines for secondary headaches, but ignore the specifics of primary headaches. Most clinicians therefore focus only on ruling out papilledema and evaluating for secondary causes without realizing that **the difficulty in headache management starts when the scan turns out to be normal!**

The sole objective while examining headache patients should be to try and distinguish the more common benign from the less common serious life-threatening causes of headache. We need to follow a flow-chart that helps us detect alarms or danger signals in patients who need further investigation. Since most headache patients have very little by way of physical findings, a good history is often the most important part of the headache consultation. If the history suggests a primary headache, then specific diagnosis is made based on pattern-recognition. Treatment will vary depending on the diagnosis for e.g. beta- blockers which are so useful in migraine will not work in cluster headache and indomethacin is the only medicine which will work in chronic paroxysmal hemicrania.

If a primary headache presents in an unusual or atypical fashion, we need to investigate and rule out the remote possibility of a secondary headache mimicking a primary headache or as we call it a **'symptomatic primary headache'**. In spite of adherence to algorithms, the headache patient seen in practice is often very different and challenging and you do come across headaches that are difficult to diagnose and treat.

4. The most useful tool for the diagnosis of headaches is a good history!

Pattern-recognition is the cornerstone of headache diagnosis. With headache patients it is important to spend more time taking the history than examining the patient. One needs to ask the right questions being aware of the entire spectrum of headaches. More than one type of headache may coexist in the same patient. Migraine may be comorbid with depression, epilepsy and stroke. Without a proper history there will be difficulty in trying to differentiate between mild or moderate migraine and tension-type headache, or in arriving at a diagnosis when the patient has more than one headache type, or an atypical presentation of a common primary headache.

History-taking is an art that improves with practice. The history should start with questions about **factors associated with the onset**. Certain triggers are known to all, but **patients are often not asked about** uncommon

triggers. **Dietary triggers** are always checked out, but they are not so common in our country. Always ask if the headaches are linked to the menstrual periods, if the patient is pregnant, if she is on oral contraceptives or on hormone replacement therapy. Ask about **sleep-related triggers** and find out if there is **simultaneous consumption of medications for other medical problems** e.g. coronary vasodilators, reserpine, hydralazine, estrogen and ranitidine, which could induce headaches.

Then there are specific questions to ask about **the head pain**. Mistakes are made by expecting that migraine head pain should always be throbbing, always unilateral. What is actually seen in practice is that migraine head pain is often generalised, and varies in character and location.[4] It is always useful to **grade the pain severity** on a scale of 0 -10. Any **change in the pattern of the headache** has to be noted**.** Knowing whether the pain comes on more at night or early in the morning on awakening helps in the diagnosis. Migraine is common on awakening in the morning and often worsens during the later part of the day. If there is a **fixed-time relationship**, and the pain comes on at the same time everyday, one must keep in mind the possibility of cluster headache.

Some patients complain of head pain that radiates to the neck and down the shoulder; mostly it is the back of the neck but sometimes on the anterior aspect of the neck also over the carotids. Rarely, the pain could start in the neck and radiate upwards to the head. Infratentorial and cervical spine pathology can refer pain to the forehead or the eye because of the convergence of C2, C3 nociceptive afferents with trigeminal afferents in the caudal trigeminal nucleus in the brainstem. In migraineurs the location of pain may fluctuate over the course of an attack and between different attacks. Some entities with **head pain may be associated with face pain** also, for e.g. facial migraine, cluster headache, cluster variants and trigeminal neuralgia. Some headaches worsen with **situational provocations** such as bending or movement or with change to the upright posture.

Sometimes while asking for **associated accompaniments** of the head pain, we remember to ask for nausea, vomiting, photophobia and phonophobia but forget to ask about the presence of autonomic features – ptosis, redness of eyes, watering of the eyes, stuffiness of nose, excessive sweating, pallor, hypotension, hypertension - these are all indirect pointers of the trigemino-autonomic cephalgias (TACs). Sometimes they may be present with some migraine headaches too. It is easier to diagnose cluster headache because of the periodicity and distinctive features, but it can be difficult to diagnose the variants. In order to distinguish these different types of headache with differing treatment, one has to ask additional questions while being aware of the different short-lasting headaches or TACs.[5]

Ask for the presence of **more than one type of headache**. Many patients are able to categorize their headaches into different types, provided they are asked the right questions. This practice of trying to clearly describe each type of headache is very important because you sometimes have to treat migraine, sinusitis-associated headache and tension-type headache in the same patient and each one of these need different drugs.

The sequence of events - where does the pain start, how does it progress and the behaviour of the patient during the attack are important. A migraine patient is irritable, prefers to lie still in a quiet dark room and does not want to talk whereas cluster headache patients would be restless, and would want to keep moving around and tension-type headache patients would be able to continue with their work. What is the **appearance of the patient during the attack**? Examination of a headache patient during the ictal phase can sometimes be more revealing than during the non-ictal phase for e.g. in an acute attack, the migraine patient may be pale, may be cold or warm, may be sweating, may manifest local oedema, temporal vessels may be distended, the scalp may be tender, he may have difficulty in speaking, you may be able to make out the associated autonomic features, there might be tenderness of muscles; in comparison, interictal examination is an anticlimax. Therefore one must always ask patients to describe an attack from start to finish.

One has to be familiar with the use of the IHS classification in routine practice. Chronic Daily Headache (CDH) and its different sub-types are the most difficult to diagnose with precision and are most often wrongly diagnosed as mixed headache syndrome. Uncommon primary headaches have also to be recognized It is **necessary to check for the IHS criteria**[1] and ask patients whether the headache worsens with continued physical activity like climbing stairs and sometimes you might get the history of worsening with bending or when the head is between the knees. Specific questioning to find out whether or not there is a **prodrome** or even a **postdrome** will give you a clue that you are dealing with migraine. You have to remember that prodromal features are different from aura. Ask how the headache clears. Is the patient left behind with a dull heaviness, does he feel drained out, does he feel fresh and how long does it take for him to come back to normal? The typical migraineur presents with unilateral throbbing headache, starts vomiting and feels better after the vomits, wants to lie down and has photophobia and phonophobia.

When you ask about age of onset and gender, you should not be surprised to see migraine in the elderly, tension-type headache in the young or cluster headache

in female patients! Although certain headaches are more common at certain ages, there are variations from the usual pattern. Migraine can present **differently at different ages**. Children may present with more vertigo and abdominal pain. Cyclic vomiting is a variant of migraine seen in the young. Adolescent females may present with basilar migraine; the chronic migraineur in her 30s or 40s may present as transformed migraine with chronic daily headache. A menopausal migraineur may present just with headache, may not vomit, and may not have photophobia and phonophobia. It helps to try and assess **the level of disability and impact of the headache on the lifestyle** of the patient, and once this is done, treatment can be fine-tuned accordingly. This is known as **Stratified Care.**

The **general medical history** and the search for underlying extra-cranial causes and systemic illness are very important in headache patients. A history of malignancy will raise the possibility of metastases, a history of vascular headaches in an individual with repeated spontaneous abortions or thrombo-embolic events should suggest the aPL syndrome and similarly it is also important to keep in mind the HIV status in relevant situations in individuals presenting with headaches. It is important to jot down the details of **previous treatment** because it helps you to know which drugs have been tried in the past so that you need not repeat the same and tells you what investigational procedures have been done earlier. It is necessary to have details of the work environment and **history of headache in other family members** which also includes children. A history of the **sleep habits** helps to know the quantum of sleep and change in the sleeping pattern. Some headaches **worsen with change of season**. Generally migraine headaches are more during summer and cluster headaches have seasonal links.

Even in a Headache Clinic, it is virtually impossible to be so perfect that you end up asking all questions to all headache patients all the time. Inspite of taking maximum care and being cautious, we still forget to ask some relevant questions. Exhaustive questionnaires make it difficult to establish rapport with the patient. In order to increase the diagnostic yield, it is always desirable not to delegate the job of history taking. As most headache patients have already seen many doctors before, one should try not to be just another doctor down the line. Unless a good rapport is established, the patient does not cooperate.

5. Not all headaches are textbook pictures.

We can diagnose headaches more effectively, if we remember that headaches are not all typical and the 3-column division between migraine, tension-type headache and cluster headache is not as clear-cut in practice as in theory. There are many atypical presentations and overlapping features. Most typical cases of migraine are seen by family physicians. As specialists we often see variations and unless we learn to recognise the variants of primary headaches, we may be mistaken in the diagnosis. You can have unilateral tension-type headache, you can have migraine without aura and migraine with aura co-existing, you can have benign intracranial hypertension without papilledema and you can have migraine which changes sites within the same attack. Migraine can present with pain in the neck, with dominant dyspeptic gastric disturbances, with autonomic features such as pallor, sweating, hypotension or hypertension, with focal neurological deficits, and with confusional states.[6] All these variations and equivalents of migraine need to be recognised. If you want to reach an earlier diagnosis in your patient who presents with headache, you have to remember that not all headaches are textbook pictures!

6. Investigation in headache patients more often tell you what it is NOT, rather than what it IS!

The next important decision is when to do a scan and whether to do a CT or an MRI scan. Since there are many advantages, an MRI scan is definitely the imaging modality of choice in headache patients with red flags or danger signals. An MR scan helps to delineate the pituitary and parasellar regions, the posterior fossa, the craniocervical regions, the facial and retropharyngeal regions and has the advantage of studying the vasculature without contrast injection. Along with the MR, one can also do the MR venogram or MR angiogram when indicated. The MR scan also helps in the differential diagnosis of other conditions that may present with headache and in those uncommon situations where the diagnosis maybe missed by doing only a CT. Plain X-rays of skull have no role in treatment and diagnosis of headache. The CT scan should be more often used for the diagnosis of sinusitis associated and rhinogenic headaches.

Lumbar puncture is not normally needed in headache patients in a chronic setting but it is important sometimes to rule out idiopathic intracranial hypertension by evaluating the CSF opening and closing pressures. The EEG is very nonspecific as far as primary headaches are concerned and the only value is in helping in the selection of anticonvulsants as prophylactics in some refractory situations. It is important to ask for investigations for 'systemic disorders' that might be associated with headaches.[7]

7. The entire spectrum of headaches is not generally known.

We often concentrate on the recognition of only migraine, tension-type headache and cluster headache. There are other primary headaches, which are less common, but definitely seen in practice, for e.g. the TACs, drug-rebound headaches, and chronic daily headache. These are usually underdiagnosed and

treated suboptimally.[3]

Cluster headache needs to be differentiated from other important entities. Primary headaches that can mimic cluster headache are supraorbital and trigeminal neuralgia, chronic paroxysmal hemicrania, the SUNCT syndrome.[5] Amongst the secondary headaches maxillary sinusitis, Tolosa- Hunt syndrome, temporal arteritis, meningiomas involving the sphenoid and petrous bones, and pituitary adenomas. A pituitary adenoma that may be impinging on the cavernous sinus can present with secondary cluster headache. Another uncommon manifestation seen with primary headaches is carotidynia. Carotidynia is pain along the course of the carotid artery in the neck and it is usually an excruciating pain with tenderness. It is seen with both migraine and cluster headache but more often with cluster headache.

Some situations may mislead into making a wrong diagnosis. The first attack of migraine if severe may appear like subarachnoid hemorrhage or acute meningitis; similarly slowly progressive secondary headaches such as idiopathic intracranial hypertension, subdural haematoma, obstructive hydrocephalus, chronic meningitis etc. can be confused with primary headaches. Sometimes secondary headaches may be present alongwith primary headaches or may mimic primary headaches or can activate latent primary headache syndromes. Rarely disorders of the nervous system that are not known to present with headaches such as demyelination or cerebrovascular disease, may present with headache and lead to confusion. CNS vasculitis and veno-occlusive disorders are often forgotten. Headaches may occur with sleep apnoea, with dementias, with Parkinson's disease and with HIV infections in the absence of space occupation. So once you know the entire spectrum then it becomes easy to spot even that rare cause of headache.

CSF pressure disorders both idiopathic intracranial hypertension and spontaneous intracranial hypotension are often not recognized. IIH can occur without papilloedema. It is important to remember that a number of systemic disorders may be associated with headache. Sound knowledge of the entire spectrum of common as well as uncommon primary and secondary headaches is one of the essential prerequisites.

8. Chronic daily headache is not the same as mixed headache!

When you are faced with patients who have headaches on a daily basis you need to know about chronic daily headaches (CDH) which comprise nearly 40% of all headache patients seen at Headache Clinics anywhere in the world. CDH has to be recognised and differentiated and the older terminology of mixed headache syndrome is no longer valid. There is no consensus on the appropriate use of the term - chronic daily headache or CDH. Some authors use it to refer to transformed migraine, others use it for any headache disorder that occurs on a daily or near daily basis regardless of the etiology and the IHS has confused the issue further by not including it in the classification. Presently, chronic daily headache can occur with chronic migraine, chronic tension type headache, new daily persistent headache, hemicrania continua; medication overuse can complicate most headaches and lead to CDH. Secondary causes of headache need to be ruled out before making a diagnosis of Primary CDH.

9. Medication Overuse Headache is an important reason for intractability and is often unrecognized!

Rebound headache due to overuse of analgesics and ergotamine also result in daily headaches but since these drugs are used to treat either migraine or tension-type headache and they convert an episodic headache to a daily headache, most of these rebound headaches are included under the heading of transformed migraine or CTTH.

Medication overuse headache has only been recently recognised but it seems to be present in epidemic proportions. Why and how they occur is uncertain, but it is likely to be due to involvement of some of the receptors in migraine patients. Typically the patient is a migraine sufferer who after some years of episodic attacks develops chronic daily headache; these chronic headaches lose some of their migrainous features and resemble a mix between migraine without aura and tension-type headache. The more analgesics or ergotamine the patient takes, the less well it works and more is needed. In addition to this predictable headache-medication cycle, patients are often depressed and experience sleep disturbances, difficulty in concentrating, restlessness and irritability. It might surprise everybody to know that in the presence of drug rebound headache, migraine prophylactic medicines prove useless.[3] It is the regularity and not the quantity which determines whether medication- induced headaches will occur. Many patients are continuously taking Ergotamine because they were started on these drugs and never went back to see their original doctor because they felt better. Another interesting point to note is that while ergotamine when used rightly for migraine or wrongly for tension-type headache can easily produce ergotamine-rebound headaches, when used for cluster headache, it has not resulted in rebound, nobody knows why and in fact ergotamine is advocated on a daily basis for the prophylaxis of cluster headache.[8] Aspirin and Acetaminophen, Codeine and Caffeine containing analgesics, Ergotamine mainly, Dihydroergotamine less commonly, and sometimes sedative hypnotics and tranquilisers, if used for a long period of time can cause rebound headaches.

10. The treatment of headache is much more than

just a prescription! And different headaches have different treatment!

The treatment of headaches is much more than just a prescription! Explanation, education and reassurance form the mainstay of therapy. The patient needs to know why the head pains, more so when his CT scan is normal. Explanations like neck muscle strain caused by fatigue or worry or stress or poor posture or a lack of exercise usually works better than telling him that nothing is wrong or that he is a neurotic. Referral to a psychiatrist may be counter productive.

Some aspects of the examination and investigation of headache patients may be overlooked. Always make sure that you palpate the head and the neck for lumps and bumps, and assess the neck mobility and look for tenderness of neck muscles. Identify trigger points, look for any thickened vessels, check the pupils and look for Horner's and always ask for a history of ptosis during the ictal phase. Auscultate for bruits over the carotids, vertebrals and orbits; percuss over the sinuses and in this era of CT, transillumination is almost completely forgotten. Do not forget to examine the temporo-mandibular joint for pain, mobility, asymmetry and for abnormal clicking. In the elderly, evaluate and rule out raised intraocular pressure because acute glaucoma can present just like migraine with headache and vomiting.[4]

There is no standard treatment for all patients with headache. The older style of 'step-care' treatment of headaches advocated first the use of simple analgesics, then combination treatment and then the use of specific antimigraine agents along with supportive treatment. In more recent times, emphasis is now laid on an assessment of disability due to the headache and the impact of the headache on the lifestyle so that right at the first step a stratified care approach is used based on a categorisation of all headaches as mild, moderate and severe. Based on an awareness of the latest treatment options, you take a decision of how aggressively you need to treat a patient. It is not realized that unless we make the perfect diagnosis of a primary headache, we will not be able to choose the appropriate treatment.

There are a number of nonpharmacological measures which again are underutilised in headache management. A number of patients for various reasons do not take the medicines prescribed and do not follow up with doctors. It could be the cost of the drugs, could be a lack of faith in the doctor and his attitude, or could be a slow rate of improvement. All these need to be explained to patients; therefore setting up a realistic level of expectation is a very important part of patient education in headache management. When the patient is not improving you have to be constantly alert to try and detect reasons for intractability; it could be a wrong diagnosis or wrong selection of drugs, there might be an ongoing rebound headache, you may have not made adjustments for some special situations, there might be a co-existing secondary headache or a systemic cause which has not been investigated, there might be an associated medical disorder or psychiatric co-morbidity, there might be need for combining nonpharmacological measures, there might be a situation where the patient will improve with inpatient hospitalization, or finally the patient may not be taking the treatment which has been prescribed. So one has to constantly reflect on the treatment administered, so as to find out why the patient is not improving.[5]

Then again there are certain situations where you have to fine tune the treatment and modify it for e.g. children, elderly and pregnant women need to be treated differently. You have to be aware that many elderly patients may be having contraindications because of other diseases, they may be using other drugs which may interact with headache medications, or some of the headache medication may have some anticholinergic or cardiac effects and an elderly might require a much smaller dosage and a more gradual build up. Certain drugs and most antimigraine drugs are contraindicated in pregnancy and breast feeding. Lastly, there are situations when headache patients need hospitalization. If the patient is in status migrainosus and continues to vomit incessantly, if there is a refractory headache or intractability on a daily basis with co-morbidity, if there is a rebound headache or when you have to wean off Ergotamine and use some substitute parenteral medication or if there are some systemic disorders then you may have to admit the patient.

Conclusion

To avoid mistakes in headache management you have to recognize the presence of underlying diseases, you need to make a check list of uncommon causes of headache, you need to recognize conditions which can mimic migraine, you need to investigate early and appropriately, you need to know all details of headache pharmacotherapy and you must employ non-pharmacological measures in addition to pharmacotherapy.

One should be familiar with the IHS classification. We need to realize that headaches are no longer just vascular and muscular, that there are now new theories for the pathogenesis, that there are now many drugs in the pipeline, that there are new routes of administration, that many acute drugs are now available as nasal sprays, that there are new concepts in headache management, that the yield of headache clinics are much more because of focus and specialisation and that in some cases inpatient management produces better, quicker results.

Ideal headache management needs correct diagnosis, right choice of medications, a sound rapport with the

patient and a compassionate nature. This article while aiming to improve the awareness of headache as a bonafide symptom which needs more respect, more education and greater emphasis on management, hopes to emphasize that Headache Medicine is a new emerging area that is on the way to becoming a credible subspeciality and should hopefully in the future be more attractive to younger clinicians.

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