You're the Flight Surgeon

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You're the flight surgeon working sick call one cold, winter morning in an overseas flight medicine clinic. Your first patient is a 27-yr-old male KC-135 pilot who is complaining of right ear pain, facial weakness, and dysgeusia, an altered sense of taste. He appears anxious and reports the ear pain, present over his right ear, started yesterday evening before bedtime. Upon waking this morning, he noticed excessive drooling from the right side of his mouth, particularly when brushing his teeth. He also thought it was strange that his cinnamon-flavored toothpaste tasted completely bland. When he examined himself in the mirror, he noticed his face appeared distorted, with visible drooping of his mouth and eyebrow on the right side. He also noted that he was unable to fully close his right eye. He denied having experienced similar symptoms in the past. He immediately called his squadron to let them know he would be unable to fly that day because he was going to sick call to be examined.

On exam, he is afebrile with normal vital signs. He is alert and oriented to person, place, and date. His speech is fluent and clear, and he is able to follow commands without difficulty. Examination of the head and his facial movement reveals drooping of the right corner of the mouth, asymmetric smile with effort, flattening of the right nasolabial fold, incomplete closure of the right eye, and no movement of the eyebrow and forehead on the right side. There are no vesicles, rash, or evidence of erythema or swelling over the face or ears, and both tympanic membranes are normal in appearance. Vision and extraocular eye movements are normal. The tongue and uvula are midline and otherwise normal in appearance. Sensation is normal across the face. Motor strength is grade 5/5 throughout both the arms and legs, and the remainder of the neurological exam is normal.

1. Given the above information, what is the most likely diagnosis at this time?

A. Ischemic or hemorrhagic stroke.
B. Ramsay-Hunt syndrome.
C. Lyme disease.
D. Bell's palsy.
E. Melkersson-Rosenthal syndrome.

ANSWER/DISCUSSION

1. D. Bell's palsy is an idiopathic, acute, unilateral, peripheral nerve palsy involving the facial nerve (CN VII). It has an annual incidence of between 11–40 cases per 100,000 persons and is found to be more common in those 15 to 45 yr old, those with diabetes, upper respiratory ailments, or compromised immune systems, or during pregnancy.[3,16] The facial nerve is a mixed nerve and has motor, sensory, and parasympathetic components. Its four major functions are: 1) voluntary facial movement via motor fibers; 2) lacrimal, submandibular, and sublingual gland secretions via parasympathetic fibers; 3) taste of the anterior two-thirds of the tongue via afferent fibers; and 4) sensation of external auditory canal and pinna via somatic afferent fibers.[9] Understanding the components that constitute the facial nerve helps illustrate the various symptoms that can result from facial nerve paralysis or paresis. These common clinical features include impaired ipsilateral movement of the affected side of the face with drooping of the eyebrow or mouth, loss of facial creases and nasolabial fold, or inability to close the eye. Other presenting symptoms may include ipsilateral earache, hyperacusis (increased sensitivity to certain frequencies and volumes of noise), tinnitus, dysgeusia, and decreased tear production.[3,14,16] The diagnosis of Bell's palsy is a clinical one and is based on the consideration that there is diffuse involvement of the facial nerve, onset is acute with a progressive course, often reaching maximal clinical symptoms within 72 h, and that recovery of some degree is present within 6 mo.[3,5]

In diagnosing the patient, perhaps the most vital distinction to make is whether you are dealing with a central (upper motor neuron) or a peripheral (lower motor neuron) pattern of facial paresis. This will assist you in ruling out stroke, which is perhaps the alarming etiology within the differential diagnosis of Bell's palsy. One is able to make this differentiation by examining the forehead—the muscles of the forehead receive bilateral innervation at the central level but ipsilateral innervation at the peripheral level.[3] Thus, a patient who has a drooping mouth but is able to wrinkle his/her forehead must be worked up immediately with stroke as the most distressing concern.[9] In our example, the patient has complete weakness of the forehead muscles and diffuse involvement of the facial nerve with no other cranial nerve involvement, and the remainder of his neurological exam is normal, making stroke an unlikely etiology.

Ramsay-Hunt syndrome refers to facial palsy that is caused by the reactivation of the varicella zoster virus within the geniculate ganglion of the facial nerve. Its clinical features include herpetiform vesicles on the pinna or pharynx, periauricular pain, and possible inner ear

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dysfunction. It has a worse prognosis than Bell's palsy and, without proper treatment, only 20% achieve complete recovery.[11] Lyme disease can also result in facial nerve palsy, which may be unilateral or bilateral. It typically occurs in younger patients and is associated with heart block, arthritis, vertigo, and hearing loss.[10] In addition to a known history of tick bite and the classic "bull's-eye" erythema migrans rash (both of which may or may not be present), features that should be considered distinctive to facial palsy caused by Lyme disease are prodromes of nontender swelling and erythema of the face.[1] Melkersson-Rosenthal syndrome is a rare condition that results in the classic triad of recurrent facial paralysis, episodic facial swelling, and a fissured tongue, although incomplete presentations are more commonly seen than the complete triad.[7] Your patient presented with no vesicles or rash seen, specifically on the face or ears, making Ramsay-Hunt syndrome unlikely. Also absent was any evidence of nontender swelling and erythema of the face. This, along with the current timeframe of occurring during the winter months, which relates to a decreased incidence of tick-borne illnesses, makes Lyme disease an improbable cause of this patient's symptoms.[4] Finally, your patient denies any history of recurrent symptoms and his tongue is noted to have a normal appearance, thereby eliminating Melkersson-Rosenthal syndrome as a likely etiology.

Using the above information and applying it to the House-Brackmann scale, you determine this patient's classification of facial nerve dysfunction to be most consistent with a grade V, or severe, dysfunction. The House-Brackmann scale is a clinical tool used to document the degree of facial paralysis, predict probability of good prognosis, and serve as an objective record of progress during recovery. It assesses gross facial features and symmetry, both at rest and during motion. The grading is from I to VI, with the former being completely normal function and the latter being total paralysis.[16] Based upon the information gathered during this evaluation, including his current House-Brackmann classification, you formulate your treatment plan using the most current evidence-based guidance.

2. Given the above information, which of the following treatment plans is most recommended?

A. Early, short-term use of oral prednisone (60 to 80 mg per day) accompanied by thorough eye care.
B. Early, short-term use of oral prednisone (60 to 80 mg per day), combined with antiviral therapy and thorough eye care.
C. Surgical decompression followed by intensive physical therapy.
D. Early use of a methylprednisolone dose pack as directed, accompanied by thorough eye care.

ANSWER/DISCUSSION

2. B. Recent literature and consensus guidelines continue to strongly recommend the use of high-dose, short-term corticosteroids (prednisone 60 to 80 mg per day) given for 1 wk and started within 72 h of symptom onset. In regard to the combined use of corticosteroids and antivirals, reviews have been less supportive; however, consensus guidelines have described the possibility for small improvements in facial palsy symptoms with low risk of harm.[3,8] In consideration of these findings, one recommendation has been to reserve antiviral use (such as valacyclovir 1000 mg three times daily for 1 wk) in combination with corticosteroids for severe cases of Bell's palsy, grade IV or higher on the House-Brackmann scale.[2] Thorough eye care for those with either incomplete or poor eye closure remains imperative to avoid corneal damage. Supportive care should include lubricating tears or ointments and eye patching at night. The literature shows no consensus for the benefit of, or indication for, decompressive surgery in the treatment of Bell's palsy. Risks associated with surgery include seizures, unilateral hearing loss, cerebrospinal fluid leak, and facial nerve injury.[16] Physical therapy has not been proven beneficial or harmful in the management of Bell's palsy.[2] Several different modalities of physical therapy have been used and include relaxation exercises, mime therapy, massage, electrical stimulation, acupuncture, heat therapy, biofeedback, and any combination of these options.[3]

You treated your patient with an early combination of corticosteroids and valacyclovir; unfortunately, his symptoms did not initially improve and physical therapy was added into his treatment plan. He was referred to Neurology for further evaluation and a noncontrast internal auditory canal magnetic resonance imaging was completed, which was read as normal. Approximately 6 mo after symptom onset, reevaluation by Neurology revealed significant subjective motor improvement and complete resolution of the dysgeusia. Interestingly, he developed new symptoms of intermittent right eye synkinetic movements associated with mouth closure, along with occasional right orbital muscle twitching. On the day of initial evaluation, the patient was very eager to start treatment and also desperate to know if and how soon it might be until he regains normal function. What would you tell him?

3. Which of the following statements regarding prognosis is FALSE?

A. Overall prognosis is related to the severity of the lesion and those with incomplete lesions tend to recover better than those with complete lesions.
B. Despite appropriate treatment, 30% of patients will be left with some type of residual symptoms.
C. The prognosis is favorable if some recovery is seen within the first 21 d of onset.
D. In relation to age, those who are older tend to show more complete recovery from their lesions in shorter amounts of time compared to those who are younger.

ANSWER/DISCUSSION

3. D. The psychological impact of facial paralysis can be significant and will vary among patients. It may be helpful, therefore, to know information about prognosis and recurrence to aid your discussion during the patient encounter. For example, being aware that even without treatment the prognosis of Bell's palsy can be good. Left untreated, 85% of patients will experience some level of improvement within 2-3 wk.[9] It is true that overall prognosis is related to the severity of the lesion; in patients with complete paralysis and no treatment, 70% will experience complete restoration of facial function within 6 mo, while in patients with incomplete paralysis, 94% will do the same. Further related to prognosis, at least 70% of treated patients will experience full

recovery within days. Unfortunately, that means approximately 30% of patients will be left with some type of residual impairment despite treatment.[3,16] In regard to recurrence, the facial palsy may recur on either the ipsilateral or contralateral side in 7–15% of patients, with a mean time to recurrence of about 10 yr. A third or fourth attack would be less likely, occurring in 3% and 1.5% of cases, respectively. Finally, it is important to know that recurrence has not been shown to indicate a worse prognosis for recovery.[13]

It has now been 7 mo since the onset of the Bell's palsy. The patient has shown unusually slow improvement in symptoms and incomplete recovery of his facial paralysis. On reevaluation he is noted to have mild smile asymmetry, weakness in holding air in the right cheek, and an inability to whistle. He displays intermittent right eye synkinetic movements with mouth closure and occasional intermittent right orbital muscle twitching was noted. He now displays complete eye closure and was able to discontinue nightly use of an eye patch. He is also able to offer resistance to forced eye opening, indicating adequate corneal protection. No communication difficulties exist. He is not in a high-performance aircraft and therefore has no need to perform an anti-G straining maneuver. He has demonstrated the ability to properly don and wear an aviator mask without inducing muscle spasms or experiencing air leakage.

4. What would be the most appropriate aeromedical disposition?

A. As the member demonstrates no functional limitations, no waiver should be required. He should be returned to flying status without further delay.
B. The member demonstrated incomplete recovery; therefore, the guidance is clear. His flying days are over.
C. A time-limited aeromedical waiver restricting the member from flying high-performance aircraft appears appropriate for recommendation to the waiver authority.
D. Refer the member to a new neurologist for a second opinion. This case seems complicated and it's better to be safe than sorry.

ANSWER/DISCUSSION

4. C. The member demonstrated incomplete recovery; however, any residual findings are not functionally limiting. Operational assessment is reassuring for use of life support equipment, there are no concerns for vision or communication complications, and the member is not required to perform anti-G straining maneuvers. According to the U.S. Air Force Waiver Guide, an isolated episode of Bell's palsy with complete recovery and no clinical or functional residua is not disqualifying and does not require a waiver. Incomplete clinical recovery or recurrent episodes of Bell's palsy are disqualifying for all flying classes and must be considered for a waiver based on the outcome of recovery and level of residual defects.[13] The U.S. Army's Aeromedical Policy Letters indicate a similar disposition in which waiver is required for any evidence of incomplete recovery, and each case will be evaluated on an individual basis.[15] A review of Navy regulations yields identical results.[12] The Federal Aviation Administration does not specifically address Bell's palsy, but does indicate that demyelinating diseases require Federal Aviation Administration decision for disposition.[6]

This particular pilot was granted a 1-yr waiver with a restriction from high-performance aircraft. An advantage of this time-limited waiver is that it allows for the potential of additional recovery. At the end of this period, the patient should be at the fullest extent of neurological rehabilitation. It also will give a time-tested period to assess how he will perform functionally. If in the future a transition to high-performance aircraft is desired, a functional assessment demonstrating adequate performance of an anti-G straining maneuver would be required.

After 1 yr the patient was reevaluated and was noted to have had essentially no change in neurological status. However, the member was able to demonstrate that he could safely and successfully perform in an operational environment, as his residual signs of neurological pathology were minimal. It was also felt that the risk for any future symptom increase or deterioration was very low and aeromedically acceptable. Due to this, an indefinite flying waiver with restriction from high-performance aircraft was recommended and approved.

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You are the flight surgeon preparing to leave your office one Friday afternoon when your boss stops by to see if you wouldn't mind doing him a small favor. Apparently the Wing commander just called and wants his 17-yr-old son checked out before he takes his Flying Class I physical at the U.S. Air Force Academy next week. The General explained that his son is extremely healthy and, in fact, "has never been sick a day in his life." He just wants the exam done as a precautionary measure, since the only thing his son has ever wanted to do was become an Air Force pilot. You graciously agree and a few moments later they arrive.

In walks a strapping young man who looks as healthy as the General walking in behind him had described. There is nothing significant in his medical history and he has no complaints. Vital signs are normal and you begin the physical exam. The head, ears, eyes, nose, and throat exam went well; however, as you auscultate the chest you hear a grade 2/6 systolic ejection murmur with a split S1 along with a split S2 that is wide and fixed.

1. What is the most likely diagnosis at this point?

A. Still's murmur.
B. Patent foramen ovale.
C. Ventral septal defect (VSD).
D. Atrial septal defect (ASD).

ANSWER/DISCUSSION

1. D. Approximately 10–15% of ASDs are diagnosed in adulthood, making it one of the most common congenital heart lesions diagnosed as an adult.[10] A systolic ejection murmur with a fixed split second heart sound that does not vary with respiration is pathognomonic for this lesion. This split is due to the increased blood flow across the pulmonic valve, resulting in delayed closure. There also may be a midsystolic murmur heard best along the upper left sternal border consistent with a right ventricular outflow murmur. A VSD murmur is a grade 2–3/6 harsh holosystolic murmur that is best heard over the left lower sternal border. A Still's murmur is an innocent murmur that is vibratory, sometimes called a "musical" murmur, and is also usually found at the left lower sternal border. It can be easily differentiated from a VSD by auscultating while the patient is supine, as it is louder in that position and diminishes when the patient sits up. Position has no effect on a VSD murmur. A patent foramen ovale usually has no distinguishable murmur associated with it.

"All done Doc?" the General asks. You explain that you hear a heart murmur on exam so you would like to obtain an electrocardiogram (EKG).

2. What classic findings if present on the EKG would help confirm your diagnosis?

A. A secondary R wave (R') in the right precordial leads (V1-3) and a wide, slurred S wave in the lateral leads.
B. ST elevation in all leads except in aVR, V1, and III.
C. Mild right-axis deviation and tall R waves in right ventricular leads with deep S waves in left ventricular leads.
D. Prolonged QRS, prominent QS in V1, and broad R waves in V5, V6, I, and aVL with deep S waves in V1-V2.

ANSWER/DISCUSSION

2. C. Mild right-axis deviation and tall R waves in right ventricular leads with deep S waves in left ventricular leads are seen with an ASD. A secondary R wave (R') in the right precordial leads (V1-3) and a wide, slurred S wave in the lateral leads is the EKG finding of a right bundle branch block. Stage 1 pericarditis shows ST elevation in all leads except in aVR, V1, and III. Prolonged QRS, prominent QS in V1, and broad R waves in V5, V6, I, and aVL with deep S waves in V1-V2 are indicative of left bundle branch block.

You inform the General that the exam and EKG could be consistent with a diagnosis of an ASD. He is a bit taken aback because no one has

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