How the Breathing Arm Syndrome is Diagnosed, the Neurophysiological Implications and its Treatment

More than a hundred years ago the Breathing Arm Syndrome was first investigated by Wilhelm Heinrich Erb, who is known for his contributions to neurology and the discovery of Erb's Palsy. The Breathing Arm Syndrome is also known as Respiratory Synkinesis of the upper arm. At the time when it was first reported, Respiratory Synkinesis was difficult to diagnose and challenging to understand as the mechanism with which the nerves regrow was unknown.

This study gauges the method with which the peripheral nervous system regenerates after the numerous ways the nerves are damaged, the interesting process to accurately diagnosing the condition, and the challenges the patients encounter. The existing literature was reviewed, and I provided insight from my experience of observing how a case of Breathing Arm Syndrome is diagnosed. This provided me with the ability to conclude that unfortunately there is no cure and very little treatment for those with the Breathing Arm Syndrome. However, one must be optimistic because, in the future, new treatments from leading laboratories around the world will mean that people affected will regain their livelihood.

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About Student text:

Introduction

In this study, I assess how one can develop the Breathing Arm Syndrome (formally known as respiratory synkinesis of the proximal arm) and analyse the neurophysiological findings, how it is diagnosed and what treatments are available.

Causation of the Breathing Arm Syndrome

The condition is caused by the respiratory synkinesis of the upper arm which involves the 'synchronous contraction of one or multiple upper limb muscles with the diaphragm muscle' (Chan Chun Kong D, Breiner A., Wolff G., 2019). However, this definition can vary slightly as it is possible for the simultaneous contraction of intercostal muscles between the ribs and the muscles of the arm. Although respiratory synkinesis is an extremely rare condition with only a handful of cases diagnosed, the Breathing Arm Syndrome is the most common form of this condition seen in the population. The synchronous contraction of the muscles in the upper arm, more specifically the bicep and deltoid. Fundamentally, The Breathing Arm is due to the breakage and regeneration of the broken nerve which reconnects to one of the respiratory nerves. The trauma to the shoulder or collar bone can be caused by intense trauma, resulting in the breakage of certain nerves in the brachial plexus (set of nerves in the shoulder that contain C5-C8 nerves leading to the arm and hand, as well as the thoracic nerve T1) causing paralysis.

Nerve Breakage

In general, the most common type of intense trauma in adolescents and adults is predominantly caused by car accidents, lacerations, gunshot wounds, or surgery. Other cases in adults have been caused by central nervous system diseases such as meningitis, syringomyelia, and syringobulbia. Bacterial meningitis can cause many peripheral and central nervous system problems due to its infection of the lining of the brain. If left untreated it can cause cranial nerve dysfunction, muscular hypertonia, and paralysis. Syringomyelia involves damage to the spinal cord caused by a syrinx (fluid filled cavity), leading to muscular atrophy (when muscle wastes away), stiffness, and pain which predominantly affects the back, shoulders, arms, and legs. This can also lead to syringobulbia, which is a similar condition to syringomyelia, where the syrinx extends to the lower brainstem. All of these diseases can create further nerve conditions by pressing on and disrupting the pathway for electrical impulses. This leads to the breakage of the nerves. The nerve disruption and breakage of the motor pathway lead to Erb's palsy which is the injury of the C5-C6 in the brachial plexus and the symptoms

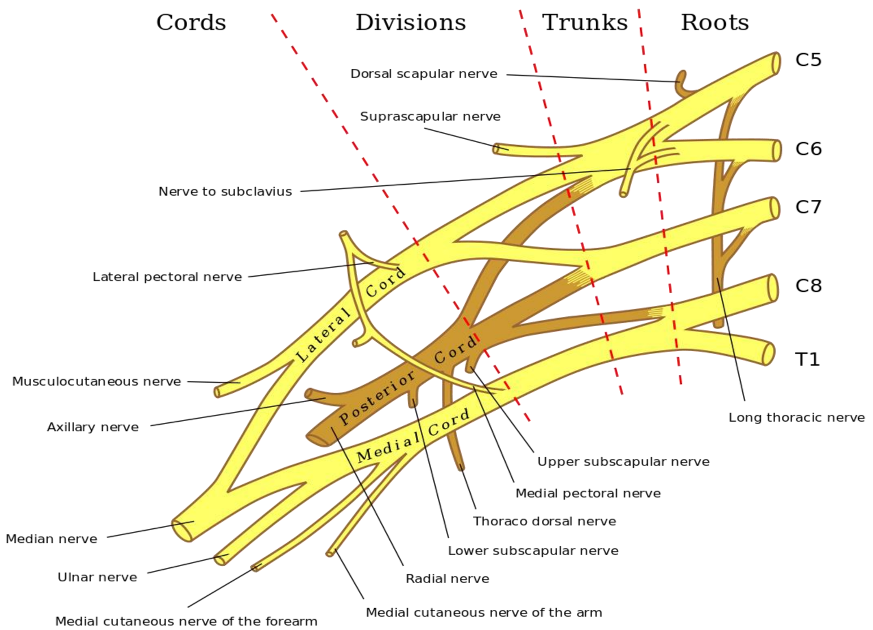

Figure 1
(Wikipedia, 2020)

can range from signs of weakness and soreness, due to muscular atrophy, to paralysis of the arm. Figure 1 shows the anatomy of the brachial plexus.

There are very rare idiopathic cases which have no apparent cause as the breakage of the nerves in respiratory synkinesis can potentially be due to neuralgic amyotrophy, which is a condition where the nerves of the peripheral nervous system experience abrupt and acute pain which leads to the degeneration of the muscles and reduced function and has a recovery time of months to years. Contrary to this, if there is a lack of reliable medical history of the prior trauma and on examination if there are no scars from intense trauma it may be classified as idiopathic. (Chan Chun Kong D, Breiner A., Wolff G., 2019)

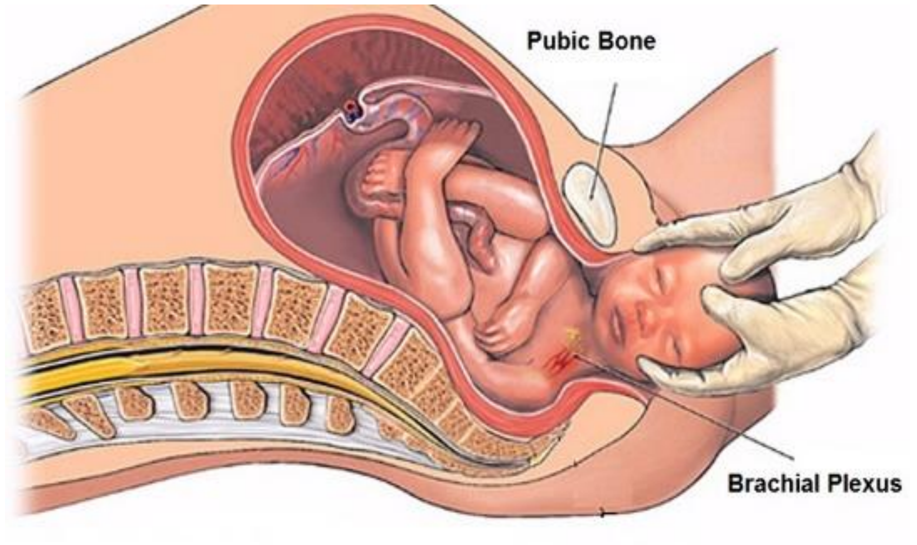

Figure 2
(Pauzillah Binti Hj, D., 2019)

Respiratory synkinesis manifests itself differently in new-borns who can develop the condition from complications during birth including dystocia (failure to exit mother's pelvis due to physical blockage) (reference). There are also numerous cases involving new-borns developing Obstetric Brachial Plexus Palsy (OBPP) which is defined as a brachial plexus nerve injury that occurs during early delivery (Pauzillah Binti Hj, D., 2019). This means that the new-born's arms will be paralysed meaning the motor pathway will be broken so the nerve cannot conduct. I have personally witnessed the diagnosis of this condition during my recent work experience placement when I saw a 59-year-old lady with a prognosis of OBPP. To confirm that she has OBPP an electromyogram (EMG) was used. An EMG is a clinical motor examination where impulse activity is recorded and analysed. A nerve conduction study is also used as part of the EMG to find how fast the impulses travel down the nerve when stimulated. The results of the test are compared with data gained from healthy individuals so that a conclusion can be made for a definitive diagnosis. In her case there was electrical activity lingering when the EMG and in her upper arm. The nerve conduction study showed that there was still a motor pathway. During the examination, the patient's inspiration and expiration were monitored synchronously with the bursts of activity that were seen when conducting the EMG test. The bursts of activity would be as the patient breathes in and then would stop as she breathes out. This is because breathing in is voluntary movement and uses conscious thought, but as she breathes out the relaxation of the intercostal muscles or diaphragm is involuntary meaning no motor units are seen. Furthermore, this encounter with respiratory synkinesis ignited a need to research more and understand how Erb's Palsy led to the discovery of the Breathing Arm Syndrome. After significant research, the first papers available online date back to 1951 (Robinson P.K., 1951). However, more than a hundred years ago the Breathing Arm Syndrome was first investigated by Wilhelm Heinrich Erb, who is known for his contributions to neurology and the discovery of Erb's Palsy.

Nerve Regeneration

In Erb's Palsy the C5 - C6 nerves in the brachial plexus are damaged causing the patient to have atrophy of the nerves, deltoid, and bicep as well as possible paralysis of the upper arm. Respiratory synkinesis is only possible due to the astounding way in which nerve cells of the peripheral nervous system regenerate themselves. However, where the break occurs, the axons in the C5 - C6 nerves start to degenerate. This is due to the nerves being extremely long cells that can reach all the way down the limb and the nucleus is very far away in the spinal cord. The first step for regrowth is for the ends of the cells to connect in some way. The C5 - C6 nerve connects to the phrenic nerve, long thoracic nerve, and the mid-cervical spinal nerves that control the diaphragm and intercostal muscles. Reconnection to the long thoracic nerve is more common as it also part of the brachial plexus and stems from all C5-C8 and T1 nerves seen in figure 1.

The regeneration occurs as the often-idle Schwann cells can de-differentiate into a stem cell state which is essential to bridging the two sides. Schwann cells are the cells seen in Figure 3 that line the axon, creating the myelin sheath which increases the nerve's conductivity making it more efficient. However, for the nerve to recover fully another cell normally used for wound healing called fibroblasts are needed, and till 2010 this use of fibroblasts was not known (How injured nerves grow themselves back – ScienceDaily, 2010). The fibroblasts can send signals to the idle Schwann cells to form a cord shape that attaches to cut ends of the axon. Also, the cords can guide the regrowth of both the axons back together across the wound. For regrowth, there is a response to the ephrin-B signal created by the fibroblasts depending on the Sox2 transcription ability. Sox2 is normally found in undifferentiated embryonic stem cells. Sox2 is also known as SRY-box2 and is needed in embryonic and neural stem cells for self-regeneration and pluripotency. New research has also found that Sox2 is one of the key ingredients in making adult cells reprogram into a cell similar to an embryonic stem cell and this is what happens to the Schwann cells. The Schwann cell's ability to move and migrate into a clump and help given to the axons are reliant on the ephrin-B signal (Parrinello et al., 2010). However, if the patient lacks healthy Schwann cells at the site of the nerve breakage, treatment is available by supplementing healthy and fully functional Schwann cells obtained from nerves prepared by an in vitro system or reinforcing the damaged nerve with Schwann cells induced with adipose stem cells. Another strategy to help regenerate peripheral nerves is to apply polymer microspheres which transport vascular endothelial growth factor to the nerve repair sites in a sustainable way. This uses the capabilities of endothelial cells that can change states like Schwann cells. The vascular endothelial growth factor helps to boost neurogenesis leading to better functional outcomes and prolongs the regeneration of the nerve. Unfortunately the latter of the two treatment methods are not ready to be used on humans as simple animal models are still being refined so that they can be evaluated fully for their effect on nerve regeneration and the potential of successfully repairing peripheral nerves.

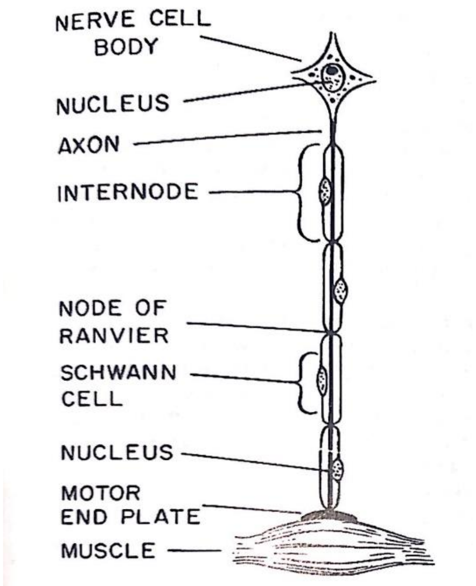

Figure 3
(Adams, Victor and Ropper, 1997, pp.1304–1305)

EMG Examination

All the patients suspected of having this condition go through a clinical motor examination. The power in the upper limb was measured using the grading recommended by the Medical Research Council so that there were no inconsistencies when examining the patients (Htut et al., 2007). Furthermore, the recovery of the shoulder and elbow functionality was measured to better evaluate the recovery of the upper limb as a whole instead of individual muscles. This was done using the Narakas' functional scoring system (Htut et al., 2007). After clinical examination, a doctor will begin an electromyogram test. An EMG is used to understand the impulses and movements of muscles. The motor neuron's electrical signal that passes through the muscles is extrapolated and is converted to waves signals and sounds that come in bursts as the muscles contract. An EMG is performed when a patient has tingling, numbness, muscle weakness, pain or cramping in the muscle, paralysis, or involuntary muscle twitching. This will help them to find any possible muscle disorders such as muscular dystrophy, motor neuron disease, or respiratory synkinesis. However, an EMG can also consist of a nerve conduction study. Using electrical stimulation pads, a current is passed through the nerve at different points along the arm. The speed at which the current passes is recorded between areas of the nerve. This means that nerve damage can be seen as it will be extremely slow and breakage in the nerve will result in their being no conduction through a particular area. This will be able to help confirm the prognosis.

Case Study

Respiratory synkinesis is only able to be seen through an electromyogram. In this case a needle with a microphone attached to the end is inserted into the bicep or deltoid muscles. When breathing in, the diaphragm and intercostal muscles contract – a voluntary action – accompanied by the bicep and deltoid contracting. When breathing out, the diaphragm and intercostal muscles relax – an involuntary action – again accompanied by the bicep and deltoid relaxing. The contraction and relaxation are seen by not only an electrical activity but also the sound it makes, picked up through the microphone. During contraction and relaxation of the bicep and deltoid in a person without respiratory synkinesis you will hear extremely loud crackles. However, for someone with the Breathing Arm Syndrome, you would only hear the crackles during the voluntary contraction but not during the involuntary relaxation. During the examination of the patient the key to knowing is telling them to take deep breaths in and out resulting in the electrical activity trace in figure 4. This is because the external intercostal muscles only contract during deep breathing.

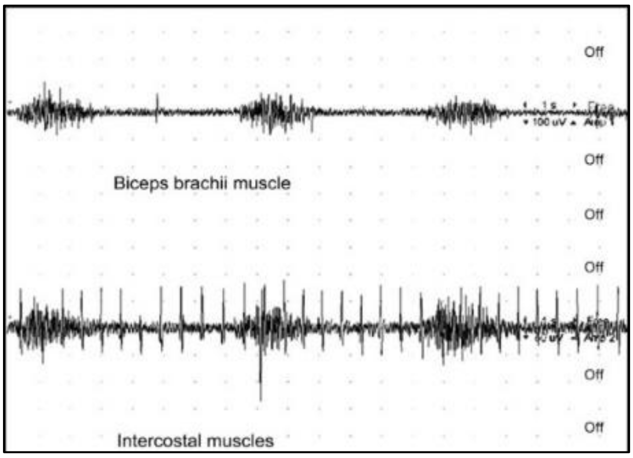

Figure 4
(Lam and Engstrom, 2010)

The patient that showed these traces was a `young man that became paralysed in the right proximal arm from a stab wound and one year later he underwent "microsurgical anastomosis" for the root of the C5 and C6 nerves (Lam and Engstrom, 2010). This surgery was performed in order to reconnect both sides of the cut axon so that regeneration can occur. Fourteen months postoperatively he had involuntary movements of his biceps and deltoid muscles synchronously with his breathing (Lam and Engstrom, 2010).

This shows the simultaneous contractions of both the 5th intercostal muscle and the bicep muscle associated with the regular bursts of activity due to the intercostal muscles during inspiration and no motor units were seen when the patient was expiring or when the patient was holding their breath. The phrenic nerve conducts the innervation impulses from the C3 - C5 nerve roots to the diaphragm showing where the impulses originate from. The regrowth of the musculocutaneous and axillary nerve in the brachial plexus connects the regenerating phrenic nerve causing reinnervation of the biceps brachii and deltoid. This results in the voluntary contraction of the biceps and deltoid coinciding with the contraction with his breathing.

Possible Treatment

After extensive research I found that there are very few treatment options available to patients with this condition, predominantly because, such rare cases are not often discussed and talked about in depth, meaning many of the neurologists and neurophysiologists are unable to diagnose respiratory synkinesis properly. When it is diagnosed after a long period of time, the root of the C5 – C6 would have receded and degenerated back to the spinal cord. Therefore, the damaged ends of the nerves that need to reconnect in order to regenerate do not, and the Schwann cell response never occurs. As research shows the spinal cord white and grey matter does not regenerate effectively, therefore surgery will not help to recover movement. For patients that are diagnosed early, there are a few possibilities, such as surgery to reconnect the C5 – C6 to its original root as well as physiotherapy afterwards to regain more movement. As I mentioned in the introduction one of the many causes for respiratory synkinesis is the surgery itself and other types of respiratory synkinesis can occur like the connection of the C7 nerve (controls the forearm's pronation, its internal rotation, its extension, and its adduction – Erb's palsy plus) as well as many more like the breathing hand syndrome.

The condition gravely impairs the quality of life for the individuals and families of those who have it. Most of the patients are often young and have their whole lives ahead of them making their situation even more tragic and due to the increasing number of road traffic incidents, these injuries are becoming more common. Unfortunately, there are very few healthcare professionals who are able to diagnose this condition because of the lack of doctors who are qualified to perform tests like an EMG. This mainly due to the specialisation needed to become qualified and due to how many extra years of training and studying are needed to become fully qualified.

From this we can sadly conclude that there is often nothing doctors can do to cure or treat certain conditions, but we know that many great things are coming in the future because of the growing research and development of embryonic and neural stem cells. Furthermore, new treatments from leading laboratories around the world will mean that people affected by Respiratory Synkinesis will regain their livelihoods.

Word Count -2719

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