Complex Regional Pain Syndrome

Body Perception Disturbance (BPD) in CRPS

Current and emerging therapeutic approaches including desensitization techniques and mirror visual feedback, together with the introduction of a new clinical tool for the early identification of BPD.

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McCabe and Lewis deserve considerable credit for this innovative contribution to the care of people with CRPS. I’ve no doubt that in the very near future all practitioners and physical therapists concerned with the plight of those with neuropathic pain will be skilled in the assessment and treatment of body perception disorders.

— Peter A. Moskovitz, MD

Complex Regional Pain Syndrome (CRPS) is a chronic pain condition of unknown aetiology that commonly occurs following trauma to a limb, although it may occur spontaneously. It is defined as type 1 or type 2, depending on whether known major nerve damage is absent or present, respectively.1,2 Patients with Complex Regional Pain Syndrome commonly describe a diverse range of sensory and motor problems. These include pain to touch or the threat of touch, temperature, colour and sweating abnormalities, problems in initiating movement and reduced function. Changes in body perception are perhaps less easily identified since patients are commonly reluctant to discuss these phenomena unless directly questioned. They often express altered perceptions, thoughts and feelings about their affected limb. They may describe their limb with negative emotional feelings such as hate and anger, disgust and repulsion.3

Patients with CRPS commonly report that the affected limb is psychologically ‘detached’ from the remainder of their unaffected body (a sense of disowning) such that it feels alien and outside of their control.3 An extreme form of detachment is expressed by some as a desperate desire to amputate the limb.3,6,7 Despite understanding clinical opinion, advising against amputation, some patients continue to express this intense urge to amputate the limb.

The purpose of this article is to help the practitioner understand BPD in CRPS by providing a theoretical understanding of body perception processes—both normal and aberrant—and how these may relate to body schema. We provide a definition of body perception disturbances and introduce a new clinical tool, The Bath CRPS Body Perception Disturbance Scale (See Appendix 1) to aid the practitioner in the identification and assessment of BPD in CRPS. Finally, we will discuss current and emerging therapeutic approaches that target central mechanisms for the resolution of BPD.

Presentation of Disturbances in Body Perception in CRPS Patients

The symptoms of CRPS include pain, usually in a single limb, together with associated unilateral color, temperature and sweating abnormalities. There may be trophic changes such as altered hair and nail growth and impairments in motor control. All of these symptoms are well described and a routine clinical investigation would encompass appropriate questions to determine their presence or absence. What a clinician may not cover is an assessment for the more subtle symptoms seen in CRPS that relate to body perception disturbance. With careful, targeted questions these symptoms are frequently found to be present.

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limb and can commonly describe, in some detail, how they plan to get rid of it. In addition, when asked where they would wish this hypothetical incision to occur, they can unhesitatingly identify the exact part of their limb where they would like this to be. Commonly this “hypothetical amputation line” forms the boundary between ownership and alienation.

Many patients report that what they see when looking at their affected limb is often at odds with how the limb feels. Subjectively, the limb may be perceived as being much larger, heavier or different in temperature or pressure from objective assessment. For example, the limb feels burning hot, yet is cool to the touch. Rather than a general distortion of the whole limb, discrete parts of the affected limb may also be perceived as grossly enlarged or missing. These sensory misrepresentations of the limb are commonly accompanied by, or perhaps contribute to, a mis-localization of the limb. Patients typically report a difficulty in knowing how their affected limb is positioned despite a heightened awareness due to pain. They describe holding their limb in what feels to be a normal and more comfortable position but are unaware that it is actually abnormal until others draw their attention to it.

Typically, patients are reluctant to look at their affected limb—choosing to position it outside of their view or covering it up in some way. The lack of visualization of the affected limb may in itself have implications on altered body perception. Commonly, patients do not wish to touch the allodynic limb and clearly avoid thinking about it. A lack of conscious attention to the limb may well contribute to the perpetuation of alterations in perception about the limb. Furthermore, patients may express a dislike of looking at the anatomically-matched limb of another and feel pain or a sense of discomfort when they do. How this BPD occurs, they can unhesitatingly identify the exact part of their limb where they would like this to be. Commonly this “hypothetical amputation line” forms the boundary between ownership and alienation.

There is a close relationship between the sensory and motor systems in order for the body to discriminate between sensory information arising from self-movement and that occurring outside of the body. Within the motor planning system, a feed forward and feedback process ensures that anticipated sensory input from a movement is matched with actual sensory feedback and any discrepancies are monitored to update future baseline assumptions. If this system becomes disrupted then actions arising from the body may be misattributed to another person or object outside of the body such as auditory hallucinations in schizophrenia. In this scenario, the individual perceives that voices are speaking to them from another when in reality their own voice box is generating the words they hear, albeit the actual sound is suppressed.

The cortical body schema has been shown to influence the typically conscious process of how people perceive their bodies. However, body perception is surprisingly malleable. Manipulation of tactile inputs in healthy individuals can create an illusion that a body part such as the nose has extended beyond what is anatomically possible. The individual perceives this dramatic distortion in body shape within seconds of tactile stimuli commencing. Similarly, we can extend the borders of our bodies simply by visualising another being touched. For example, it has been shown that our own body maps become activated—in an anatomically precise manner—when we visualise another being touched. Further, we perceive the same emotions that this action of touch may evoke in the other individual. This ‘mimicking’ within our own bodies of actions and emotions that we are passively observed occurring in others, is thought to be facilitated via the mirror neurone system. Mirror neurones are thought to be created through an integrated sensorimotor experience—such as the observation of an action—and can be modified over time. What is still hotly debated is whether mirror neurones evolve purely via sensorimotor experience and are crucial to it, whether we are born with mirror neurones to facilitate action understanding, or if it is a combination of both theories (See Heyes 2009 for a review). What is clear, however, is that the mirror neurone system enables us to closely relate to experiences that happen outside of our own bodies and perceive these experiences so accurately we can feel the same emotions these external actions evoke. This makes the mirror neurone system a potentially powerful tool for therapeutic interventions but, by the same token, something that clinicians should consider when they are communicating and working with patients. The clinician’s own body movements may generate pain and emotional consequences simply by the patient observing them.

The perception of one’s own and that of other human bodies has been shown to occur in a discreet visual processing region
termed the “extrastriate body area” or EBA. Increased activity occurs in this region when we view human bodies or body parts rather than faces or objects. However, the level of activity in the EBA is similar whether we are viewing our own body, someone who is familiar to us or someone who is unfamiliar. This suggests that the EBA is not able to discriminate between different human bodies but is purely involved in body recognition (see Minnebusch et al., 2009 for review). In addition to the EBA, the “fusiform body area” (FSA) has also been described as a center for body recognition and shown to be active on recognition of the whole body rather than specific body parts. Finally, it has been hypothesised that a “body-identification network” exists which links the EBA and FSA to a region in the right inferior parietal cortex. This network recognizes both whole bodies and body parts and may share some commonalities with similar networks for face recognition.

Underlying Mechanisms of Body Perception Disturbance in CRPS

We have already described how body perception evolves from a complex interaction of central and peripheral processing. The patients’ reports of disturbances in limb perception of their CRPS-affected limb suggest that the body-identification network has become disrupted so that distorted mental images and thoughts become associated with the way the brain represents the affected limb or body schema. We will consider here how such aberrant perceptions may arise.

Recent brain imaging studies have provided evidence of cortical reorganization in CRPS. Several of these studies have found shrinkage in the cortical representation of the affected hand in the primary (SI) and secondary (SII) somatosensory cortex. Given that this cortical region is one area known to be responsible for the body schema, these changes support the hypothesis that the body schema is disrupted in CRPS. In addition, SI also provides information on spatial localization and perception. With altered body representation and spatial perception, limbs may become difficult to locate and may understandably feel alien if they now seemingly exist outside the sufferer’s revised spatial field. Without visual feedback, body parts not contained within revised spatial fields or represented within internal body maps may start to lack definition and “disappear” (see McCabe et al., 2009). Changes have also been reported in parietal function in patients with CRPS.

As the parietal cortex is important in the activation and maintenance of an internal representation of a desired movement, altered sensory perception may occur in the presence of disturbed motor planning. Although misattribution of ones own actions to another has not been described in CRPS, certainly patients have described that their limbs seemingly move without their knowledge or control. In addition, the parietal cortex is a central part of the proposed body-identification network and perhaps patients with CRPS become alienated from their own limbs, and that of others, due to poor recognition of body parts. It would be interesting to see if facial recognition is also affected in this group due to the close relationship between body and face recognition processes.

The fact that patients stop looking at their limbs and are distrest by visualising the limbs of others suggests to us that sensory input has become altered and the mirror neuron system is active. What we do not know is whether these changes are the cause or affect of altered BPD. Careful prospective observation of those with CRPS may help us to understand the timeline of onset of these changes and how they relate to other BPD and changes in the cortical maps.

Body Perception Disturbances in CRPS and How to Assess

Body perception disturbances in CRPS are defined as the individual’s perceived alteration of their CRPS affected body part while regarding the remainder of their body as normal. These perceived alterations include one or more of the following components:

- Distortions in shape and/or size, weight, pressure or temperature that are different to objective assessment
- Loss of specific anatomical parts during mental visualization
- Hostile feelings
- A sense of disowning the body part
- Impaired limb position sense
- A desire to amputate

Since these bizarre feelings and thoughts are often difficult for the patients themselves to make sense of, they are typically reluctant to disclose them to others. Health professionals can broach the subject by asking general questions such as “how do you feel emotionally about your affected limb?” and “does your limb feel as if it belongs to you?” These questions can form the starting point for discussion in order to gauge the presence and extent of BPD. It is important that the clinician stresses that these perceptions are entirely normal in CRPS and that the patient is not hallucinating or thought to be fabricating symptoms.

To gain a better insight into how the patient perceives their limb, the clinician can ask the individual to visualise a mental picture of their affected limb with their eyes closed. It may help to first ask them to describe their unaffected limbs, starting with those most distal to the affected limb. For example, in a left upper limb-affected patient you would ask them to first compare and contrast their lower limbs with each other. Ask them to consider if the feet, knees and hips are perceived as the same size and shape as each other, the legs the same length. Take each section of the limb in turn so that the patient really learns to attend to each part of that limb and become aware of any perceived difference between the limbs. You would then move their attention to their right upper limb and finally the left, affected limb. Invite them to compare and contrast the size and shape of their two limbs and consider if all sections of those limbs are present, what size or shape they perceive them to be and if the limbs are the same length. Typically patients describe vivid distortions in shape and size of the limb and some are unable to visualise specific anatomical parts such as the shoulder or forearm. These alterations in shape and size can be particular to specific limb regions. For example, the forearm can be perceived as elongated and thin and the thumb foreshortened and much fatter. Patients may describe a “mitt like” appearance of the hand so that the fingers are merged into a single, shapeless clump with the thumb isolated. They are typically surprised when undertaking this mental visualization task for the first time as patients may be unaware that they perceive their limb in such a distorted manner. These simple visualization techniques probably give us access to an unconscious representation of the affected limb in the mind and appear to be the clinical correlates of the cortical reorganization described above.
The Bath CRPS Body Perception Disturbance Scale

The Bath CRPS BPD Scale, presented in Appendix 1, provides a comprehensive assessment of the extent to which body perception disturbance is experienced and offers a means by which changes in body perception can be monitored over time. Based on previous research, this tool has been developed clinically to identify patients with disturbances in body perception in order to target rehabilitation appropriately.

In order to capture the nature and extent of these perceptual disturbances, the scale has seven items that cover different aspects identified through previous research. Items 1-4 and 6 rate individual aspects of body perception disturbance on a 0 to 10 scale to establish the extent of perception abnormality. Item 5 determines the subjective perception of changes in size, weight pressure and temperature. The final item illustrates the mental representation of both limbs using the contralateral unaffected limb as a comparator. The patient is asked to close their eyes and generate a mental picture commencing with their unaffected side first. The assessor draws a pencil line picture of both limbs as the participant describes their mental image. The resultant drawing can then be assessed by the patient for accuracy and amended accordingly. The drawing is independently graded to determine the extent of mental representation disturbance. In addition to rating changes in subjective body perception, the advantage of this final item is that it provides a visual quantification of perceived distortions of the affected limb which can be compared over different time points particularly over the course of rehabilitation.

Scoring of the Bath CRPS Body Perception Disturbance Scale

The sum of numerical ratings from items 1 to 4 and 6b are added to items 5 and 6a (scored no=0 yes=1). The mental representation drawing is graded on a three-point scale: no distortion=0 distortion=1, severe distortion=2. If either a distortion in size or shape is depicted within the drawing or the accompanying textual descriptions—i.e., not anatomically consistent with the actual size or shape of the limb—the rating ‘distortion’ is given. If one or more segments of the limb are missing it is rated as a ‘severe distortion.’ A sum total gives an overall score and a higher score denotes greater disturbance, with 57 being the maximum total score.

Treatment Approaches

The general principle in addressing body perception disturbance is to provide corrective input to normalize attitudes, perceptions and emotions about the affected limb. A combination of sensory, motor and cognitive inputs targeted at normalizing the affected limb body schema is key. Encouraging patients to look at and touch the affected area, where tolerated, provides corrective sensory inputs to accurately update the body schema. Cognitive strategies that encourage patients to think about their affected limb in a more positive way—such as being a useful part of their body once more—may help to reinforce a sense of limb ownership. Importantly, thinking, looking and touching the affected limb should be undertaken for short periods as frequently as possible in order to maintain corrective updating of the body schema. These guiding principles can be incorporated into many specific interventions in body perception disturbance rehabilitation.

Mirror Visual Feedback (MVF)

MVF, summarized in Appendix 2, has become a useful aid in CRPS rehabilitation due to the increasing body of evidence demonstrating its pain-relieving benefits (see Ramachandran et al, 2009 for review). Although not proven, it would also seem reasonable that MVF could have a positive impact on body perception disturbance with the mirror providing a corrective visual representation of the affected limb and thereby contributing to a normalization of the body schema. Patients have reported that they are more willing to look at their affected limb as a consequence of MVF suggesting greater ownership and improved emotional attachment to that limb.

Ramachandran and Rogers-Ramachandran first described the use of MVF when they set out to relieve paralysis and spasm in amputees. Their rationale for this approach was based on the theory that involuntary movements and paralysis in a phantom limb arise from a combination of pre-amputation memories and a mismatch between motor output and sensory feedback. They hypothesized that, by ‘giving back’ the amputated limb via a mirror illusion, they would provide, albeit artificially, the anticipated sensory feedback. This would help suppress involuntary movements or enable free movement of a previously paralyzed phantom limb. Through regular use of mirror visual feedback (10 weeks of 10 minutes visualization per day) subjects described a relief of their phantom pain and, in some cases, a total cessation of their phantom limb pain.

Of the eight subjects who participated in the first pilot study of mirror visual feedback (MVF) for CRPS Type 1, five had a marked reduction in pain at rest and upon movement six weeks after regular use of the device and the remaining three reported no benefit. There was no relief of pain on movement when both limbs were visualised or a non-reflective surface viewed. Disease duration appeared to be a significant factor in this study as all those who reported an analgesic benefit had symptoms less than one year compared to the non-responders of 2-3 years. Since this study was conducted, further research, including those with chronic disease duration, has confirmed the analgesic properties of MVF in CRPS.

What has also become apparent over time is that MVF can exacerbate symptoms for some patients and therefore careful screening is required. Data from our clinical practice (publication in preparation) has demonstrated that when patients cannot execute simultaneous bilateral, symmetrical movements with the affected and unaffected limbs, then pain and other sensory disturbances will rapidly increase in the affected limb. In addition, those who cannot believe in the illusion find little benefit from MVF. This is in line with research in those with amputee phantom limb pain that has shown that belief in the visual illusion is required for analgesic benefit to be achieved. Our data suggests that those who cannot perform bilateral synchronised movements have higher levels of pain, greater BPD and different patterns of sympathetic response to standardized stimuli. Further work is required to precisely define this phenotype of CRPS but clinicians should be alert to the fact that MVF is not always innocuous. Patient information should make it clear that MVF should be discontinued if symptoms are exacerbated or bilateral movements cannot be achieved (see Appendix 2).
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Notes on Administration of the Bath CRPS Body Perception Disturbance Scale

**Administration**

The CRPS body perception disturbance scale is used to capture changes in self-perception of the affected limb. Based on previous research, this tool has been developed clinically to identify patients with disturbances in body perception in order to target appropriate treatment interventions. In order to capture the nature and extent of these perceptual disturbances the questionnaire has seven items that cover different aspects as identified through research.

Participants are asked to rate five of these aspects on a 0 to 10 scale to establish the extent of abnormality within that item. The sixth item determines the subjective perception of changes in size, weight pressure and temperature. The final item illustrates the mental representation of both the affected and unaffected limbs.

The participant is asked to close their eyes and generate a mental picture of both the affected and matching unaffected limbs commencing with their unaffected side first. During the description the investigator draws a picture of both limbs as the participant verbally describes their mental image. The resultant drawing is then assessed by the participant for accuracy. The drawing is independently graded on a three-point scale; no distortion=0, distortion=1, severe distortion=2. If either a distortion in size or shape is depicted within the drawing or the accompanying textual descriptions, i.e. that it is not anatomically consistent with the actual size or shape of the limb, the rating ‘distortion’ is given. If one or segments of the limb are missing this is rated as a ‘severe distortion’.

A sum total gives an overall score and a higher score denote greater disturbance with 57 being the maximum total score.

Appendix 1: The Bath CRPS Body Perception Disturbance Scale*

Developed by Jennifer S. Lewis, The Royal National Hospital for Rheumatic Diseases
Bath, England. v2. ©2008. All rights reserved.

Patient name ________________________ Date ________ Assessment no. 1 2 3 4 5
Diagnosis ___________________________ Date of symptom onset __________
Body part affected: 1)_________________________
2)_________________________
3)_________________________
1) On a scale of 0-10 how much a part of your body does the affected part feel?
Very much a part = 0__ 1__ 2__ 3__ 4__ 5__ 6__ 7__ 8__ 9__ 10 = Completely detached
2) On a scale of 0-10 how aware are you of the physical position of your limb?
Very aware = 0__ 1__ 2__ 3__ 4__ 5__ 6__ 7__ 8__ 9__ 10 = Completely unaware
3) On a scale of 0-10 how much attention do you pay to your limb in terms of looking at it and thinking about it?
Full attention = 0__ 1__ 2__ 3__ 4__ 5__ 6__ 7__ 8__ 9__ 10 = No attention
4) On a scale of 0-10 how strong are the emotional feelings that you have about your limb?
Strongly positive = 0__ 1__ 2__ 3__ 4__ 5__ 6__ 7__ 8__ 9__ 10 = Strongly negative
5) Is there a difference between how your affected limb looks or is on touch compared to how it feels to you in terms of the following:
   Size yes □ no □ Comment __________________________
   Temperature yes □ no □ Comment __________________________
   Pressure yes □ no □ Comment __________________________
   Weight yes □ no □ Comment __________________________
6a) Have you ever had a desire to amputate the limb?  Yes □ No □
6b) If yes, how strong is that desire now?
Not at all = 0__ 1__ 2__ 3__ 4__ 5__ 6__ 7__ 8__ 9__ 10 = Very strong
Desired amputation site __________________________
7) With eyes closed describe a mental image of your affected and unaffected body parts (drawn by assessor during patient description then verified by the patient)

This is an accurate account of my image of my affected body part.
Signature ___________________________ Date __________________

*Notes on Administration of the Bath CRPS Body Perception Disturbance Scale

Scoring

The sum of numerical ratings from items 1 to 4 and 6b are added to items 5 and 6a (scored no=0 yes=1). The mental representation drawing is graded on a three-point scale; no distortion=0 distortion=1, severe distortion=2. If either a distortion in size or shape is depicted within the drawing or the accompanying textual descriptions, i.e. that it is not anatomically consistent with the actual size or shape of the limb, the rating ‘distortion’ is given. If one or segments of the limb are missing this is rated as a ‘severe distortion’.

A sum total gives an overall score and a higher score denote greater disturbance with 57 being the maximum total score.
Appendix 2. Patient Guidelines for the Use of Mirror Visual Feedback in Complex Regional Pain Syndrome

Note: these guidelines should only be used following consultation with your clinician.

What is mirror visual feedback?
Mirrors have been found to be of use in the treatment of amputee phantom limb and we too have found that some patients with Complex Regional Pain Syndrome (CRPS) also report a reduction in their pain. It is thought that the mirror works by ‘tricking’ the brain to believe that the painful limb is now ‘normal’, by looking at the reflection of the unaffected limb. This helps, over time, to correct the messages to the brain and thereby reducing pain that, in turn, may assist exercise.

What should I do?
Mirror visual feedback should become part of your planned exercise program. Follow the instructions below 4-5 times a day (or as directed) but only use the mirror for brief periods (maximum 10 minutes) or until you feel you are no longer able to concentrate. It is best to use it a short time and often. Like any new technique, it will need practice and may need several trials before you are used to it.

• Find a quiet room where you will be able to concentrate and not be interrupted.
• Ensure that you are seated/lying comfortably with your back well supported.
• Position the mirror so that the reflective surface is facing your unaffected limb and your affected (the one with CRPS) limb is hidden behind the mirror (Figures 1 & 2).
• Lean forward so that you are now able to see the mirror image of your unaffected limb in the mirror and simply look at this reflection without moving either your affected or unaffected limb. Concentrate hard on the mirror reflection for a minimum of a couple of minutes.
• Once you are comfortable with this, very slowly start to move both limbs in exactly the same way while still looking at the reflection in the mirror. You may want someone with you the first time you do this to ensure you are moving both limbs at the same time and in the same way. It does not matter what movements you chose to do as long as you do the same with both limbs. However, there may be specific exercises that your Physiotherapist has prescribed for you that you could perform while using the mirror.
• If pain prevents movement of your affected limb, then try to imagine that you are moving it in the same manner as the unaffected limb.
• If pain or stiffness becomes a problem while you are exercising, stop and go back to simply looking at the mirror image while both limbs are resting.

Important: do not use the mirror for prolonged periods as your concentration levels will drop and always move your affected and unaffected limbs in the same way.

Are there any side effects with this treatment?
Occasionally, when using the mirror, one may feel unusual sensations in the limb that is hidden from view. For example:
• heavier or lighter,
• warmer or colder
• reduced sensation or as if it is ‘floating’
• or even that you have an additional limb.

All of these sensations are much less common when both limbs are moving in the same manner and should last no more than a few seconds or minutes after you have stopped using the mirror. If at any time you find a sensation uncomfortable or disturbing then simply stop the particular exercise and view both limbs without the mirror and the sensation should quickly pass.

Desensitization
Desensitization could be considered as providing corrective tactile input to the affected limb body schema. Traditionally this has been conducted using a range of textured fabrics but more recent research suggests that electrical sensory discrimination training devices may also play a role. Shown to provide pain relief in those with phantom limb pain, this powerful visual technique has great potential for further exploration in those with CRPS.

More radically, the use of neuromodulation techniques—such as Motor Cortex Stimulation which directly target cortical representation and have been shown to relieve pain—may also influence body perception. Clearly
this is not a first choice therapy but we would suggest that both peripheral and central mechanisms may need to be targeted for effective reversal of body perception disturbance. Alternatively, it is possible that BPD naturally reverses as other CRPS symptoms resolve.

Conclusion
Disturbance in body perception is common and is a normal finding in CRPS. It is frequently overlooked, or not considered, by the clinical team since patients are reluctant to discuss them due to a fear of being disbelieved or considered psychologically unsound. An understanding of BPD and how they present in CRPS is crucial for effective rehabilitation and therapies targeted at the underlying mechanisms. In order for us to learn more about these distressing perceptions, we would suggest that every therapeutic trial includes an assessment of BPD to see how they change over time and how they are influenced by different treatment modalities.

References