

**THE MANY FACES OF PHANTOM VISION:
CHARLES BONNET SYNDROME (CBS)**

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**I HAVE NO FINANCIAL INTERESTS
TO DISCLOSE**

THE STORY

IN 1769, CHARLES BONNET, A SWISS NATURALIST AND PHILOSOPHER, REPORTED THAT HIS GRANDFATHER, CHARLES LULLIN, EXPERIENCED WELL-FORMED VISUAL HALLUCINATIONS IN THE ABSENCE OF ANY KNOWN PSYCHOPATHOLOGY OR COGNITIVE IMPAIRMENT.

BRIEFLY, MONSIEUR LULLIN, AT AGE 89 (11 YEARS AFTER CATARACT SURGERY), BEGAN TO SEE ASTONISHING IMAGES OF MEN, WOMEN, CARRIAGES, AND BUILDINGS.

THE STORY

THE FIGURES APPEARED IN MOVEMENT: APPROACHING, RECEDING, BECOMING LARGER OR SMALLER, DISAPPEARING THEN REAPPEARING. BUILDINGS WOULD RISE IN FRONT OF HIS EYES, SHOWING THEIR EXTERIOR CONSTRUCTION. TAPESTRIES IN HIS APARTMENT WOULD CHANGE AND BECOME THOSE OF A "RICHER TASTE." AT OTHER TIMES, THE TAPESTRIES WOULD BE COVERED IN PAINTINGS OF SCENIC VIEWS.

THE STORY

THESE VISUAL IMAGES APPEARED WITH HIS FULL KNOWLEDGE THAT THEY WERE NOT ACTUALLY PHYSICALLY PRESENT.

THE TERM CHARLES BONNET SYNDROME (CBS) WAS COINED IN 1967 BY ANOTHER SWISS SCIENTIST – GEORGES DE MOSSIER.

CRITERIA FOR CBS

- VISUAL HALLUCINATIONS
 - ♦ FORMED
 - ♦ COMPLEX
 - ♦ PERSISTENT OR REPETITIVE
 - ♦ STEREOTYPED

CRITERIA FOR CBS

- INSIGHT IS FULLY OR PARTIALLY RETAINED
- ABSENCE OF PRIMARY OR SECONDARY DELUSIONS
- ABSENCE OF HALLUCINATIONS IN OTHER MODALITIES
- DESCRIBED AS “WATCHING A MOVIE WITH NO SOUND”

CRITERIA FOR CBS

- LESS CLEAR WHETHER PHOTOPSIAS WITH FLICKERING AND FLASHING LIGHTS SEEN BY PATIENTS WITH ADVANCED AMD ARE PART OF OR DIFFERENT FROM CBS
- HOWEVER, GEOMETRIC SHAPES AND PATTERNS ARE PART OF CBS

KEY CLINICAL FEATURES

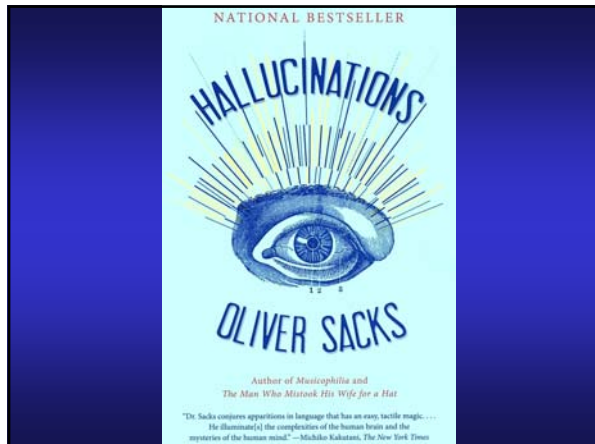
- GENDER
- VISIONS ARE FORMED, COMPLEX, AND ELABORATE; REMARKABLY CRISP AND DETAILED
- THE VISIONS TEND TO BE PROLONGED IF NOT CONTINUOUS
- SCENES AND THEMES TEND TO BE ANIMATED AND COLORFUL

KEY CLINICAL FEATURES

- MINIATURE SPECTRES (LILLIPUTIAN FIGURES, ETC) ARE PERCEIVED BY PATIENTS AS AMUSING AND DELIGHTFUL
- INSIGHT INTO THE UNREALITY OF THE VISIONS IS EITHER PRESENT FROM THE BEGINNING OR RAPIDLY ACQUIRED; THUS, IT IS MORE OFTEN A “PSEUDOHALLUCINATION” THAN A “TRUE” HALLUCINATION

KEY CLINICAL FEATURES

- RARELY UNPLEASANT HALLUCINATIONS
- THE MOVEMENTS DO NOT TELL A STORY
- OCCASIONALLY RELATED TO PERSONAL MEMORIES BUT NOT USUALLY



- DR. SACKS DEDICATES THE FIRST CHAPTER OF HIS BOOK TO THE CBS
- MULTIPLE, UNIQUE PATIENT EXPERIENCES WITH CBS

SACKS, *HALLUCINATIONS*. NY:VINTAGE BOOKS, 2012

SOME UNUSUAL OBSERVATIONS IN CBS:

- A. "TEXT HALLUCINATIONS"
- B. "LIKE BALLOONS IN A COMIC STRIP"
- C. ANOMALIES OF COLOR, DEPTH, TRANSPARENCY, MOTION, SCALE, AND DETAIL
- D. DAZZLING, INTENSE COLOR; FINENESS AND RICHNESS OF DETAIL; EXOTIC DRESS, HATS, ETC

SACKS, *HALLUCINATIONS*. NY:VINTAGE BOOKS, 2012

DANIEL ffYTCHÉ OF LONDON

- NEURAL BASIS OF VISUAL HALLUCINATIONS
- BRAIN IMAGING STUDIES SHOW "A STRIKING CORRESPONDENCE" BETWEEN THE PARTICULAR HALLUCINATORY EXPERIENCE OF EACH PATIENT AND THE PARTICULAR PORTIONS OF THE VENTRAL VISUAL PATHWAYS IN THE VISUAL CORTEX THAT WERE ACTIVATED

SACKS, *HALLUCINATIONS*. NY:VINTAGE BOOKS, 2012

- THERE IS CLEAR DISTINCTION BETWEEN NORMAL VISUAL IMAGINATION AND ACTUAL HALLUCINATION
- THE V4 AREA OF THE VISUAL CORTEX IS NOT STIMULATED BY IMAGINED COLOR BUT IS STIMULATED BY COLORFUL HALLUCINATIONS

SACKS, *HALLUCINATIONS*. NY:VINTAGE BOOKS, 2012

- THEREFORE, "NOT ONLY SUBJECTIVELY BUT ALSO PHYSIOLOGICALLY, HALLUCINATIONS ARE UNLIKE IMAGINATION AND MUCH MORE LIKE PERCEPTION"

SACKS, *HALLUCINATIONS*. NY:VINTAGE BOOKS, 2012

- “ONE OF THE DEFINING CHARACTERISTICS OF CBS HALLUCINATIONS IS THE PRESERVATION OF INSIGHT, THE REALIZATION THAT A HALLUCINATION IS NOT REAL”

SACKS, *HALLUCINATIONS*. NY:VINTAGE BOOKS, 2012

OTHER CLINICAL FEATURES

- 20% OF CASES ARE UNDER 60 YEARS OF AGE BUT OTHERWISE HIGH MEAN AGE
- TRIGGERS COULD BE LOW ILLUMINATION, DECREASED SOCIAL CONTACT (SENSORY REDUCTION OR ISOLATION)
- IMAGES DISAPPEAR WHEN EYES ARE CLOSED OR WITH BLINKING AND WITH OCULAR SACCADES BUT NOT WITH SMOOTH PURSUIT MOVEMENTS

OTHER CLINICAL FEATURES

- PATIENTS WITH BILATERAL DISEASE, WORSE VISION, AND OLDER AGE HAD HIGHER FREQUENCY OF CBS
- CANNOT BE VOLUNTARILY INVOKED OR THE CONTENT INFLUENCED

OTHER CLINICAL FEATURES

- PREVALENCE OF 1%-40% REPORTED IN PTS WITH DECREASED CENTRAL VISION
- CBS IS NOT RECOGNIZED GENERALLY AS AN EARLY MARKER FOR DEMENTIA

CHARACTERISTICS OF CBS

- HALLUCINATIONS
- SIMPLE GEOMETRIC SHAPES
- COMPLEX SHAPES WITH RECOGNIZABLE FORMS (FACES, COMMON OBJECTS)
- TESSELLOPSIA (REGULAR, OVERLAPPING PATTERNS)

Kester, Charles Bonnet syndrome: case presentation and literature review. *Optometry*. 2009;80:360-366

CHARACTERISTICS OF CBS

- PROSOPOMETAMORPHOSIA (FACIAL DISTORTION)
- DENDROPSIA (BRANCHING FORMS)
- HYPERCHROMATOPSIA
- POLYOPIA (MULTIPLE FORMS OF ONE IMAGE)
- MICROPSIA
- MACROPSIA

Kester, Charles Bonnet syndrome: case presentation and literature review. *Optometry*. 2009;80:360-366

CHARACTERISTICS OF CBS

FREQUENCY

DAILY	27
WEEKLY	30
MONTHLY	35
LESS OFTEN	8

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

CONTENT

PEOPLE

ADULTS	80
FACES	70
CHILDREN	35
LARGE GROUPS	12
MINIATURE PEOPLE	3

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

CONTENT

ANIMALS	38
PLANTS/TREES	25
BUILDINGS	15
SCENES	15
OTHER OBJECTS	42

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

RELATIONSHIP TO SURROUNDINGS

FITTING IN WELL	22
RANDOMLY PROJECTED	37
VARIABLE	28

PRESENCE OF COLORS

ALWAYS	63
SOMETIMES	10
ONLY BLACK/WHITE	27

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

INTRINSIC MOVEMENT

ALWAYS	30
SOMETIMES	17
NEVER	53

RECURRING OBJECTS

ALWAYS	22
SOMETIMES	18
NEVER	60

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

DURATION

1-5 s	13
5-60 s	17
1-60 min	53
>1 h	15
NOT CERTAIN	2

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

FAMILIARITY OF CONTENT

ALWAYS	4
SOMETIMES	25
NEVER	65
NOT CERTAIN	2

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

INFLUENCE OF EYELIDS

ONLY WITH OPENED EYES	67
ONLY WITH CLOSED EYES	13
VARIABLE	20

STEREOTYPED HALLUCINATIONS

ALWAYS	10
SOMETIMES	5
NEVER	85

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

MOVING WITH EYES

ALWAYS	18
SOMETIMES	15
NEVER	52
NOT CERTAIN	13

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

CHARACTERISTICS OF CBS

CLARITY

AS REAL OBJECTS	27
CLEARER	45
LESS CLEAR	20
VENABLE	8

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

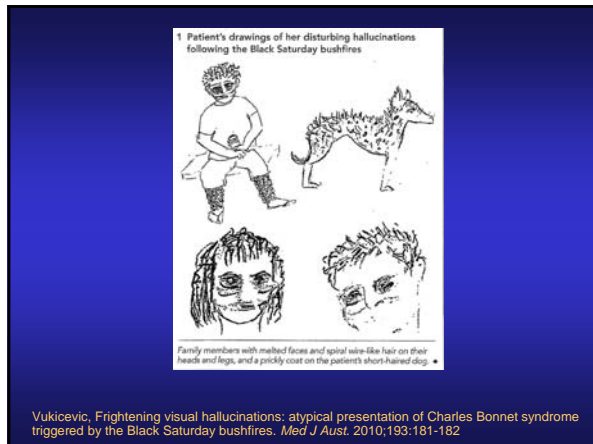
CHARACTERISTICS OF CBS

MOVEMENT "EN BLOC"

ALWAYS	22
SOMETIMES	3
NEVER	72
NOT CERTAIN	3

Teunisse et al, Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet*. 1996;347:794-797

- RARELY, AN EMOTIONALLY STRESSFUL EXPERIENCE MAY TRIGGER MORE FRIGHTFUL HALLUCINATIONS



PATHOPHYSIOLOGY OF CBS

- "IRRITATIVE" CNS LESION VS. SENSORY DEPRIVATION OR (COGAN) "RELEASE PHENOMENON"
 - ♦ "PHANTOM PAIN" ANALOGY
 - ♦ CBS WITH ENUCLEATION

MUSICAL EAR SYNDROME (MES)

- CONDITION IN PEOPLE WITH HEARING LOSS WHO SUBSEQUENTLY DEVELOP AUDITORY HALLUCINATIONS
- ALSO ASSOCIATED WITH MUSICAL HALLUCINATIONS, IN WHICH AN INDIVIDUAL HEARS MUSIC OR SOUNDS WITHOUT AN EXTERNAL SOURCE
- COMPARABLE TO CBS, AND IT HAS BEEN SUGGESTED IT COULD BE INCLUDED UNDER THE DX OF CBS

DIFFERENTIAL DIAGNOSIS

- NEUROLOGIC DISORDERS
 - ♦ HEMISPHERIC LESIONS
 - ♦ PARKINSON'S DISEASE
 - ♦ LEWY-BODY DEMENTIA
 - ♦ BRAINSTEM LESIONS
 - ♦ EPILEPSY
 - ♦ MIGRAINE
 - ♦ NARCOLEPSY-CATAPLEXY SYNDROME

DIFFERENTIAL DIAGNOSIS

- PSYCHIATRIC DISORDERS
 - ♦ ACUTE PSYCHOSIS
 - ♦ DELIRIUM
 - ♦ SCHIZOPHRENIA
 - ♦ AFFECTIVE DISORDER
 - ♦ CONVERSION REACTIONS

DIFFERENTIAL DIAGNOSIS

- TOXIC AND METABOLIC DISORDERS
 - ♦ METABOLIC ENCEPHALOPATHIES CAUSED BY CARDIOPULMONARY INSUFFICIENCY, UREMIA, HEPATIC DISEASE, ENDOCRINE DISTURBANCES, VITAMIN DEFICIENCY STATES, AND INFLAMMATORY AND INFECTIOUS DISEASES
 - ♦ HALLUCINOGENIC AGENTS
 - ♦ DRUG AND ALCOHOL WITHDRAWAL
 - ♦ MEDICATION OR TOXIN SIDE EFFECTS

DIFFERENTIAL DIAGNOSIS

- MISCELLANEOUS CONDITIONS
 - ♦ SENSORY AND SLEEP DEPRIVATION
 - ♦ HYPNOPOMPIC OR HYPNAGOGIC STATES
 - ♦ INTENSE EMOTIONAL EXPERIENCES (STRESS, GRIEF)

PATHOLOGY IMPLICATED IN CBS

- **AMD**
 - CATARACT
 - GLAUCOMA
 - MACULAR HOLES
 - LASER PHOTOCOAGULATION
 - MACULAR TRANSLOCATION SURGERY

PATHOLOGY IMPLICATED IN CBS

- CRAO
- RETINAL DETACHMENT
- OPTIC NEURITIS
- RETINITIS PIGMENTOSA
- DIABETIC RETINOPATHY
- POST- ENUCLEATION

PATHOLOGY IMPLICATED IN CBS

- ★ OPTIC PATHWAY UP TO OCCIPITAL CORTEX
- GIANT CELL ARTERITIS
- MULTIPLE SCLEROSIS
- “BLACKPATCH DELIRIUM”
- “CATARACT PSYCHOSIS”
- EVEN SIMPLE EYE PATCHING CAN DO IT

TREATMENT

- REASSURANCE THAT THE CBS IS BENIGN AND DOES NOT LEAD TO PSYCHIATRIC DISEASE
- SYMPATHY FROM CARE PROVIDERS, INCREASED SOCIAL INTERACTION, AND IMPROVED HOME LIGHTING
- TREATMENT FOR AMD LESION
- LOW-VISION THERAPY

TREATMENT

- PHARMACOLOGICAL Rx HAS HAD ONLY SPOTTY SUCCESS
 - ♦ ⊕ CARBAMAZEPINE
 - ♦ ⊕ VALPROIC ACID
 - ♦ ⊕ YI-GAN SAN (YGS) – JAPANESE HERBAL MEDICINE
 - ♦ ⊖ LEVETIRACETAM

CONCLUSION

- HETEROGENEOUS NATURE OF CBS
- GENTLY INQUIRE IF THEY SEE IMAGES THAT THEY KNOW ARE NOT THERE
- PATIENTS ARE RELUCTANT TO VOLUNTEER CBS SYMPTOMS BECAUSE OF FEAR OF BEING LABELED AS HAVING A MENTAL DISORDER

CONCLUSION

- IN OUR EXPERIENCE, PATIENTS WITH LESS THAN 5/200 VA MORE FREQUENTLY HAVE CBS (NOT PROVEN)

CONCLUSION

- ALWAYS KEEP IN MIND OTHER POTENTIAL ISSUES WITH ELDERLY PATIENTS
 - ♦ COGNITIVE FALL-OFF
 - ♦ DEMENTIA OF VARIOUS LEVELS
 - ♦ ALZHEIMER'S
 - ♦ STROKES AND MINISTROKES
 - ♦ INFLUENCE OF MEDICATIONS
 - ♦ ETC.