Acalculia

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Introduction

This article includes discussion of acalculia, acquired dyscalculia, anarithmetia, agraphic acalculia, alexic acalculia, aphasic acalculia, and spatial acalculia. The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

Overview

Calculation ability represents a complex cognitive process. It has been understood to represent a multifactorial skill, including verbal, spatial, memory, and executive function abilities. Calculation ability is frequently impaired in cases of focal brain pathology, especially posterior left parietal damage, and dementia. Acalculia is also common in posterior cortical atrophy. Contemporary neuroimaging studies suggest that arithmetic is associated with activation of specific brain areas, specifically the intraparietal sulcus; language and calculation areas are partially overlapped and partially independent. Acalculia recovery is variable and depends on different factors, such as the extension and etiology of the brain pathology.

Key points

• Calculation ability represents a multifactorial skill including verbal, spatial, memory, and executive function abilities.
• Calculation disturbances are usually observed in cases of posterior left parietal damage.
• The intraparietal sulcus has been proposed to represent the most crucial brain region in the understanding and the use of quantities.
• A major distinction can be established between primary and secondary acalculia.
• Acalculia is most often caused by a stroke, tumor, or trauma and it is usually present in dementia.

Historical note and terminology

Henschen introduced the term “acalculia” to refer to the impairments in mathematical abilities in patients with brain damage (Henschen 1925). Berger distinguished 2 different types of acalculia: primary and secondary acalculia. Secondary acalculia refers to calculation defects resulting from a different cognitive deficit: memory disorders, attention impairments, language defects, spatial deficits, etc. (Berger 1926). Gerstmann proposed that acalculia is observed together with agraphia, disorders in right-left orientation, and finger agnosia, representing the basic brain syndrome usually known as “Gerstmann syndrome” (Gerstmann 1940). Gerstmann syndrome has been associated with left angular gyrus damage (Vallar 2007). It has been proposed that Gerstmann syndrome represents a disorder in the spatial representation of the body-scheme and mental rotations occurring after left parietal damage (Gold et al 1995).

Hecaen and colleagues distinguished 3 major types of calculation disorders: (1) alexia and agraphia for numbers, (2) spatial acalculia, and (3) anarithmetia (Hecaen et al 1961). Alexia and agraphia for numbers represent calculation disturbances resulting from difficulties in reading and writing quantities. Spatial acalculia represents a disorder of spatial organization where the rules for setting written digits in their proper order and position are not followed; spatial neglect and number inversions are frequently found in this disorder. Anarithmetia corresponds to primary acalculia. It implies a basic defect in computational ability.

Boller and Grafman considered that calculation abilities can be disrupted as a result of: (1) inability to appreciate the meaning of the number names, (2) visuospatial defects that interfere with the spatial arrangement of numbers and the mechanical aspects of mathematical operations, (3) inability to recall mathematical facts and appropriately use them, and (4) defects in mathematical thinking and in understanding underlying operations (Boller and Grafman 1985).

A general cognitive model of number processing and calculation has been proposed by McCloskey and colleagues (McCloskey and Caramazza 1987; McCloskey et al 1991). A distinction is drawn between the number processing
system, which comprises the mechanisms for comprehending and producing numbers, and the calculation system, which encompasses processing components required specifically for carrying out calculation. In the case of brain pathology, these components can be dissociated. Facts (eg, the multiplication tables), rules (eg, \( n \times 0 = 0 \)), and procedures (eg, the multiplication process goes from left to right) are included as elements of the calculation system. Errors in calculation observed in brain-damaged and normal subjects can result from inappropriate fact retrieval, misuse of arithmetical rules, and procedural errors.

Ardila and Rosselli have proposed a classification of acalculias (Ardila and Rosselli 2002). A basic distinction between anarithmetia (primary acalculia) and acalculia resulting from other cognitive defects (secondary acalculias) is included. Secondary acalculias may result from linguistic defects (oral or written), spatial deficits, and frontal-type disturbances, particularly perseveration, memory, and attention impairments. However, there is a certain degree of overlap among the acalculia subtypes proposed by Ardila and Rosselli. Thus, in anarithmetia some spatial deficits can be observed. Spatial acalculia associated with right hemisphere pathology is also partially an alexic acalculia (eg, some inability to read complex numbers as a result of left hemineglect can be observed). Even though arithmetical calculation is a rather complex cognitive activity requiring the participation of many elements, brain damage may result in a relatively restricted disorder. Rosca reported a patient with preserved arithmetic facts but impaired procedural knowledge (Rosca 2009b). Klessinger and colleagues studied a 56-year-old retired university professor suffering a vascular lesion in the left middle cerebral artery territory with extensive damage to perisylvian temporal, parietal, and frontal cortices, resulting in right hemiplegia, severe aphasia, and apraxia of speech. Regardless of the severe language disturbance and his difficulties with processing both phonological and orthographic number words, he demonstrated largely intact algebraic fact knowledge, procedures, and conceptual knowledge; this case illustrates that some aspects of mathematical processing can be preserved despite severe disruption to the language ability (Klessinger et al 2007).

Hittmair-Delazer and colleagues described a patient affected by an inability to recall and use “arithmetical facts” of single-digit multiplication and division (Hittmair-Delazer et al 1994). This impairment contrasted with the preservation of a wide range of complex notions (cardinality judgments, recognition of arithmetical signs, written calculations, solving arithmetical problems, additions and subtractions). This patient's difficulty stemmed from an inability to monitor the sequence of operations that calculation procedures specified. Dehaene and Cohen described 2 patients with pure anarithmetia, 1 with a left subcortical lesion and the other with a right inferior parietal lesion and Gerstmann syndrome (Dehaene and Cohen 1997). The patient with the subcortical lesion suffered from a selective deficit of rote verbal knowledge (eg, arithmetical tables), whereas the semantic knowledge of numerical quantities was intact. The patient with the inferior parietal lesion suffered from a category-specific impairment of quantitative numerical knowledge, with preserved knowledge of rote arithmetical facts. Domahs and colleagues reported a patient with left frontal lesions due to a cerebrovascular disorder presenting defects in simple multiplication when problems were given in a mixed operations list (multiplication, addition, and subtraction) but not when the same multiplications were presented in isolation (a kind of “task-switching acalculia”) (Domahs et al 2011). Tanaka and colleagues described an unusual case of “abacus-based acalculia” (Tanaka et al 2012). Abacus users typically manipulate a mental representation of an abacus. The authors evaluated a patient who was a good abacus user and transiently lost her “mental abacus” after a right hemispheric stroke involving the dorsal premotor cortex and inferior parietal lobule.

Troiani and colleagues proposed that numerical quantifier understanding (which requires magnitude processing, for example, “at least three”) and logical quantifier understanding (which can be comprehended using a simple form of perceptual logic, such as “some”) depend on different brain neural networks; whereas numerical quantifier understanding depends on a lateral parietal-dorsolateral prefrontal network, logical quantifier understanding depends on a rostral medial prefrontal-posterior cingulate network (Troiani et al 2009). It has been further proposed that the numerical processing and calculation system includes 4 independent elements: quantitative numerical knowledge, numerical recodification, qualitative numerical knowledge, and calculation. In cases of brain pathology, each component may be impaired selectively without affecting the others, indicating that each one has different brain representation.

**Clinical manifestations**

**Presentation and course**

A major distinction can be established between primary acalculia (anarithmetia) and secondary acalculia (aphasic acalculia, alexic acalculia, agraphic acalculia, frontal acalculia, and spatial acalculia).
Anarithmetia. Anarithmetia represents a basic defect in the computational ability. Patients with anarithmetia present with a loss of numerical concepts, inability to understand quantities, defects in using syntactic rules in calculation (eg, “to borrow”), and deficits in understanding numerical signs. They may count and conserve some numerical knowledge but fail in comparing numbers, performing arithmetical operations, and solving numerical problems. The failure in calculation tasks has to be found in both oral and written operations.

Luria emphasized the defects in spatial conceptualization underlying the primary acalculia observed in left-parietal damaged patients and also emphasized the strong association between acalculia and semantic aphasia (Luria 1966). He proposed that left parietal-temporal-occipital damage could produce components of spatial apraxia, spatial agnosia, semantic aphasia, and acalculia. Luria considered that the same cognitive defects were present in semantic aphasia and acalculia; therefore, acalculia was associated with semantic aphasia.

Ardila proposed that left angular gyrus (Gerstmann) syndrome should be restated to include acalculia, finger agnosia, right left disorientation, and semantic aphasia. A single underlying deficit can account for the simultaneous presentations of these 4 clinical signs (Ardila 2014).

Aphasic acalculia. Dahmen and colleagues studied calculation deficits in patients with Broca and Wernicke types of aphasia (Dahmen et al 1982). Using a factor analysis, they were able to identify 2 different factors: numeric-symbolic and visual-spatial. The milder calculation defects found in Broca aphasia patients are derived from linguistic alterations, whereas with Wernicke aphasia, defects in visual-spatial processing significantly contribute to calculation difficulties. Syntax of calculation is impaired in Broca aphasics. These patients present morphologic errors (eg, 14 is read as 40), and they have difficulties in counting backward and in successive operations. Omission of words is noted when transcoding tasks are given to the patients (that is, converting numbers in their written representation; for example, “1245” to “one thousand two hundred forty-five”; or the inverse task).

Wernicke aphasics present semantic errors in the reading and writing of numbers. Verbal paralexias for numbers are abundant. Verbal memory defect is evident in mathematical problem solving when the patient has to retain different elements of the problem. Lexicalization (eg, 634 is written 600304) and decomposition errors are frequent (eg, 1527 is written 15 27).

In conduction aphasia both oral and written calculation may be impaired. Sequencing operations, solving of mathematical problems, and transcoding tasks are abnormal. Order and hierarchy errors may be observed. As a matter of fact, the neuroanatomical site of damage most commonly noted in conduction aphasia (left parietal lobe) lies close to the area suspected for anarithmetia (angular gyrus). Hence, it is not infrequent to find conduction aphasia associated with primary acalculia.

Calculation disturbances are also observed in other types of aphasia. In extrasylvian (transcortical) motor aphasia the patient may have difficulties in initiating and maintaining numerical sequences. Problem solving ability may be significantly impaired. In extrasylvian (transcortical) sensory aphasia, significant calculation defects are usually found. Temporal-parietal damage results in a variety of language disturbances and significant calculation defects.

Alexic acalculia. The calculation deficits observed in alexia without agraphia (occipital alexia or pure alexia) are limited to reading numbers. Defects in reading numbers parallel the defects found in reading written words and sentences. Digit-by-digit reading may be observed. There is an asymmetrical reading, and the initial digits in a number are easier to read than the last ones. The patient may read the initial digits and omit the remainders (eg, 746 is read 74). Written arithmetical operations may be difficult due to the visual exploration disorders.

Central (parietal-temporal) alexia includes an inability to read written numbers and numerical signs. The ability to perform mental calculation usually is considerably better. Often, central alexia is associated with anarithmetia. Reading and writing difficulties plus computational disturbances indicate severe acalculia.

Agraphic acalculia. Difficulties in writing numbers are correlated with the particular type of agraphia. Broca aphasia patients present a nonfluent agraphia with perseveration and order reversal. Difficulties in transcoding tasks, substitutions, and omissions are evident. Writing number sequences, particularly in reverse order, may be especially difficult.

In Wernicke aphasia, a fluent agraphia is observed. Lexical errors are frequent; abundant numerical verbal paralexias and paragraphias are observed. Language-understanding defects impair the ability to write quantities under dictation.
Some degree of apractic agraphia is frequently found in conduction aphasia. This apractic agraphia will also be found in writing quantities. Self-corrections and approximations are found, and frequently the patients fail in converting the numbers they hear in a correct graphic form.

**Frontal acalculia.** Patients with damage in the prefrontal areas of the brain may display serious difficulties in mental operations, successive operations (particularly backward operations, such as 100 minus 7), and multistep numerical problems. Written arithmetical operations are notoriously easier than mental operations. Difficulties in calculation tasks in these patients correspond to 3 different types: (1) attention difficulties, (2) perseveration, and (3) impairment of complex mathematical concepts. Attention difficulties result in defects in maintaining the conditions of the tasks and impulsiveness in answers. Perseveration results in the incorrect answer (eg, 100 minus 7: 93, 83, 73, etc.). Impairment in the use of complex mathematical concepts results in an inability to analyze the conditions of numerical problems and to develop an algorithm for its solution. Most significant defects are found in solving numerical problems, whereas elementary arithmetic is usually much better preserved. When arithmetical problems are presented in mixed formats (eg, multiplications and additions), patients with frontal damage may fail, whereas no defects are usually observed when a single operation is used and no switching is required (Domahs et al 2011).

**Spatial acalculia.** Spatial acalculia is most frequently observed in right hemisphere pathology (Ardila and Rosselli 1994; Benavides-Varela et al 2014). Hemineglect, topographic agnosia, constructional apraxia, and general spatial defects are usually correlated with spatial acalculia. Patients with spatial acalculia perform much better in orally presented arithmetical tasks than in written ones. Writing numbers is defective, with exclusive use of the right side of the page. Iterations (eg, 44 is written 444), inability to maintain a straight line, and spatial disorganization are observed. Alignment of the numbers in columns is severely abnormal. In performing written operations there may be confusion about where to locate the "carried" quantity; left neglect may result in failure to complete the operation, or inability to read the numerical sign, usually located at the left. In multiplication procedures, these patients may know the required steps in the operation, that is, when and how to carry, but they do not know where to place the quantities. These patients may have difficulty remembering multiplication tables and frequently mix procedures (eg, when subtracting, they add). Of note, patients with right hemisphere pathology more frequently make transcoding errors involving zeros because transcoding zeros entails visuospatial and integrative processes typically associated with the right hemisphere (Benavides-Varela et al 2016).

**Prognosis and complications**

The severity of acalculia and the pattern of recovery are variable. Recovery may be related to the size of the lesion and the etiology of the damage. In general, recovery in primary acalculia is limited. In secondary acalculias, recovery correlates with the recovery observed in the primary defect (aphasia, alexia, etc.). Caporali and colleagues suggest that recovery from acalculia is possible in the first months poststroke, that initial severity does not significantly influence recovery, and that initial severity correlates with recovery of auditory comprehension (Caporali et al 2000). Bernal and colleagues reported a patient with a severe acalculia and compared the fMRI pattern of activation with 4 normal age-matched controls (Bernal et al 2003). Both patient and controls showed specific task-related activation (especially left posterior temporal-parietal cortex) with some differences that may indicate brain plasticity, even though no significant recovery in calculation abilities was observed in the patient.

Basso and colleagues (Basso et al 2005) analyzed the co-occurrence of aphasia and acalculia in 98 left-brain-damaged patients and the spontaneous recovery from acalculia in 92 patients. A significant association between aphasia and acalculia was observed although 19 participants exhibited aphasia with no acalculia and 6 acalculia with no aphasia. The authors concluded that dissociations between language and calculation disorders are not uncommon. A significant improvement was observed between a first examination carried out within the first 5 months post-onset and a second examination carried out on average 5 months later.

**Clinical vignette**

A 68-year-old woman with a high school level of education was hospitalized due to a sudden loss of sensitivity in her left hemibody. At the incoming neurologic exam, left hyposthesia in face and arm, left homonymous hemianopia, disorientation in time and place, and left hemineglect were found. CT scans revealed an ischemic lesion involving the temporal and parietal branches of the right middle cerebral artery.

Neuropsychological testing indicated a significant left neglect. Neglect was observed in drawing, reading, writing, and
A severe spatial acalculia was noted. The patient could not align numbers in columns. Adding and multiplying were impossible because the patient could not find where to place the "carried" or "borrowed" quantities. She confused "plus" and "multiplication" signs and could not read numbers at the left (e.g., 9231 was read as 31). Due to these defects, the patient failed in performing all the written numerical operations that were presented. Nonetheless, her ability to perform arithmetical operations orally and to solve numerical problems was nearly normal. A diagnosis of spatial acalculia associated with spatial alexia, spatial agraphia, hemispatial neglect, and general spatial defects was presented.

During the following weeks neglect mildly improved. Still, she continued to be unable to perform any written arithmetical operation. Defects in spatial orientation continued to be significant. One year later, she began to attend a rehabilitation program that lasted for about 1 year. Improvement was significant. At the end of this program, the patient could perform simple written arithmetical operations.

**Biological basis**

**Etiology and pathogenesis**

Acalculia is most often caused by a stroke, tumor, or trauma. Acalculia is usually present in dementia (Park et al. 2013). Acalculia is also common in posterior cortical atrophy (Kas et al. 2011), corticobasal degeneration (Pantelyat et al. 2011), left posterior peri-insular infarct (Bhattacharyya et al. 2014), and left thalamic damage (Jensen 2010).

Primary acalculia is associated with left posterior parietal damage (Ardila and Rosselli 2002). Pure global acalculia has been reported following left subangular lesions (Martory et al. 2003). It has been suggested that different cerebral pathways may be responsible for processing rote numerical knowledge (e.g., multiplication tables) and semantic knowledge of numerical quantities. Dehaene and Cohen have proposed that a left subcortical network contributes to the storage and retrieval of rote verbal arithmetical facts, whereas an inferior parietal network is dedicated to the mental manipulation of numerical quantities (Dehaene and Cohen 1997). Asada and colleagues suggested that acalculia is associated with a defect in manipulating mental images; moreover, mental imagery disturbances can be found in cases of right and left parietal damage, and hence acalculia can be due to both right and left hemisphere pathology (Asada et al. 2014). “Primary progressive acalculia” as initial manifestation of a degenerative disease has been described (Ardila et al. 2003).

Neuroimaging techniques (e.g., fMRI) have been used to analyze the pattern of brain activation during calculation tasks. It has been demonstrated that different brain areas are active during arithmetical tasks, but the specific pattern of brain activity depends on the type of test that is used. It has been found that at least the following brain areas become activate during calculation: upper cortical surface and anterior aspect of the left middle frontal gyrus; supramarginal and angular gyrus (bilaterally) (Rueckert et al. 1996); left dorsolateral prefrontal and premotor cortices, Broca area, and inferior parietal cortex; and left parietal and inferior occipitotemporal regions (lingual and fusiform gyri) (Rickard et al. 2000). The diversity of brain areas involved in arithmetical processes support the assumption that calculation ability represents a multifactor skill, including verbal, spatial, memory, body knowledge, and executive function abilities (Ardila and Rosselli 2002). Dehaene and colleagues, however, have proposed that the human ability for arithmetic is associated with activation of specific brain areas (Dehaene et al. 2004). Neuroimaging studies with humans have demonstrated that the intraparietal sulcus is systematically activated during diverse number tasks and could be regarded as the most crucial brain region in the understanding and the use of quantities (Ota et al. 2009). Other brain areas such as the precentral area and the inferior prefrontal cortex are also activated when subjects engage in mental calculations. An unusual case of acalculia following an infarct restricted to the left intraparietal sulcus was reported (Ashkenazi et al. 2008); as expected, the patient presented significant deficits in basic numerical processing tasks. Defects were also observed in numerical comparisons, counting, and subitizing (direct perceptual apprehension of the numerosity of a group). The authors concluded that the underlying deficit referred to an impairment in the perception and manipulation of quantities. Using fMRI, a cluster of neurons selective for visually presented numbers in healthy human adults was found (Grotheer et al. 2016). This visual number form area was observed in the inferior temporal gyrus of both hemispheres.

Roscia proposed that arithmetic knowledge impairments can be due to either "memory" defects or "monitoring" problems. Patients with "memory" disturbances typically have left parietal lesions, whereas "monitoring" impairments...
are associated with frontal damage (Rosca 2009a). Arithmetical procedural impairments, on the other hand, can be observed in cases of basal ganglia pathology. In consequence, it can be assumed that a fronto-parieto-subcortical circuit is responsible for complex arithmetical processing.

Symptoms of Gerstmann syndrome (acalculia with agraphia, disorders in right-left orientation, and finger agnosia) (Gerstmann 1940) can be found during direct cortical stimulation in the angular gyrus region (Roux et al 2003). Using fMRI it has been observed that the left angular gyrus is not only involved in arithmetic tasks requiring simple fact retrieval, but may show significant activations as a result of relatively short training of complex calculation (Delazer et al 2003).

Colvin and colleagues (Colvin et al 2005) investigated numerical abilities in a split-brain patient using experiments that examined the hemispheres' abilities to make magnitude comparisons. One experiment examined the ability to enumerate sets of stimuli and another 2 experiments required judgments about 2 concurrently presented stimuli that were either identically coded (ie, 2 Arabic numerals, 2 number words, or 2 arrays of dots) or differently coded (eg, an Arabic numeral and a number word). Both hemispheres were equally able to enumerate stimuli and to make comparisons between numerical representations regardless of stimuli coding. However, the left hemisphere was more accurate than the right when the task involved number words. Pu and colleagues, using electrical stimulation during brain surgery, observed that different brain sites in the left parietal lobe were specifically related to multiplication or subtraction processing whereas in the right hemisphere no brain sites associated to numerical processing were found (Pu et al 2011).

Cohen and colleagues reported a patient with a lesion in the left perisylvian area who showed a severe impairment in all tasks involving numbers in a verbal format, such as reading aloud, writing to dictation, or responding verbally to questions of numerical knowledge. In contrast, her ability to manipulate nonverbal representation of numbers, ie, Arabic numerals, was comparatively well preserved (Cohen et al 2000). This observation supports the proposal that language and calculation disorders can be dissociated. When patients with aphasia are tested with a calculation battery adapted to their language difficulties, remarkable improvement in arithmetical skills may be observed (Rosca 2010).

Some authors have assumed that language and numerical concepts are differently organized in the brain and follow distinct developmental patterns in children (Gelman and Butterworth 2005). Other authors have suggested that calculation and language are mediated by partially different and also partially overlapped brain systems (Baldo and Dronkers 2007). Semenza and colleagues have emphasized that calculation and language usually share the same hemisphere (Semenza et al 2006). Using electrostimulation mapping it has also been observed that language and calculation areas are partially overlapped and partially independent (Roux et al 2009).

**Differential diagnosis**

Acalculia represents a significant component in global cognitive disorders, usually observed in dementia and in cases of traumatic brain injury. It has been proposed that mathematical abilities represent excellent predictors of general intellectual performance in dementia (Rosselli et al 1998; Remy et al 2004). Acalculia correlates with severity of dementia. Although calculation and numeral transcoding deficits are often prominent in early courses of the disease, deficits in semantic processing and basic number processing are less severe. The retrieval of arithmetic facts is generally spared with aging but can be impaired in Alzheimer disease patients. Using several mathematical tests Crutch and Warrington observed different patterns of calculation disorders in Alzheimer disease and frontotemporal dementia (Crutch and Warrington 2001). The group with frontotemporal dementia was subdivided into those with a semantic dementia and those with prominent frontal features (nonsemantic dementia). On the quantity facts test, the semantic dementia subgroup was more impaired than both the Alzheimer disease patient group and the nonsemantic frontotemporal dementia subgroup. A further analysis revealed several interesting patterns of performance, including a dissociation between impaired performance on the quantity facts and number operations tests and preserved performance on the Graded Difficulty Arithmetic Test. Of note, calculation defects in dementia can be different depending on the characteristics of the writing system; thus, it has been observed that Chinese-speaking patients make significantly more intrusion errors than English-speaking ones, due to the ideographical nature of both Chinese characters and Arabic numbers (Ting et al 2016).

Calculation abilities are strongly correlated with educational level. People with lower education may have difficulties in performing some arithmetical tasks.
Diagnostic workup

A few models of testing for calculation abilities have been developed (Deloche et al 1994; Ardila and Rosselli 2002; Denburg and Tranel 2003; Villain et al 2015). Usually it is considered that testing for acalculia should include the following tasks:

- Counting forward and backward. Patients are asked to count from 1 to 10 or from 30 to 40 (forward condition), and from 10 to 1 or 40 to 30 (backward condition).
- Writing and reading numbers with different levels of complexity (e.g., 7, 31, 106, 1639, 10002). Different quantities are dictated to the patient: 1-digit numbers, 2-digit numbers, numbers requiring the use of a positional value, etc.
- Transcoding from a verbal to a numerical code and from a numerical to a verbal code. Numbers using a numerical code are written (e.g., 7, 23, 109), and the patient is asked to write them with letters (e.g., seven, twenty-three, one hundred nine). Numbers using a verbal code are written (e.g., three, forty-nine, three hundred seventy-six) and the patient is asked to write these quantities with numbers (e.g., 3, 49, 376).
- Reading and writing arithmetical signs (+, -, ·, /, x, =), interpreting arithmetical signs (e.g., 9 + 5 = _, 9 - 5 = _).
- Magnitude comparisons: The patient is asked which number is larger and which is smaller (e.g., 79 and 103).
- Mental calculation: adding, subtracting, multiplying, and dividing single-digit simple quantities and complex (2 or 3 digits) quantities.
- Written calculation: adding, subtracting, multiplying, and dividing single-digit simple quantities, and complex (2 or 3 digits) quantities.
- Successive arithmetical operations requiring the patient to add or to subtract or to do both (e.g., 1, 4, 7,...; 100, 93, 86...).
- Aligning numbers in columns: a series of numbers (e.g., 27, 2, 2407) are dictated to the patient. The patient is required to place them in a column, as if for adding.
- Numerical problems: problems requiring the use of 1 or several numerical operations are presented to the patient (e.g., "There are 18 books in 2 shelves; in one of them there is double the number than in the other. How many books are there in each one?").
- Numerical knowledge (e.g., "How many days are in a week?" "How many weeks are there in a year?").

Ardila suggested that due to new technological advances (e.g., the extended use of pocket calculators), new assessment procedures more in accordance with current living conditions are to be developed (Ardila 2013).

Management

Several case studies have addressed the rehabilitation of calculation defects. Girelli and colleagues reported significant improvement following a rehabilitation program in 2 patients with a selective deficit in multiplication facts (Girelli et al 1996). Hittmair-Delazer and colleagues reported a patient unable to recall and use “arithmetical facts.” After the application of a rehabilitation program, a selective improvement for each single arithmetical fact was observed (Hittmair-Delazer et al 1994). Rosselli and Ardila reported the rehabilitation of a 68-year-old patient presenting with spatial acalculia associated with spatial alexia and spatial agraphia (Rosselli and Ardila 1996). The rehabilitation program was based on overcoming the hemispatial neglect and the associated spatial difficulties. A significant improvement was observed.

Tsvetkova proposes a multistep rehabilitation program for primary calculation disturbances associated with brain pathology (Tsvetkova 1996). It includes multiple activities that range from simple to progressively more complex: counting real objects, matching the quantity of object with the written number, dividing the objects in subgroups (equal and nonequal), counting the objects in the subgroups, matching the written number, training on the positional value of the numbers (units, tens, hundreds, etc.), using numbers in columns, training on arithmetical operations using a multistep and piecemeal detailed process, dividing big numbers into smaller ones, analyzing the components of the
quantities, and training in the correct sequence of arithmetical operations. Tsvetkova reports significant improvements after long-term training.

A review of the literature published from 1980 to 2007 dealing with the treatment efficacy of calculation disorders found few studies that tackled the issue of efficacy; efficiency of acalculia therapy seems promising, but data are scanty (Basso et al 2011). Rosselli and Ardila point out that rehabilitation must start with an analysis of the errors presented by the acalculic patient and of the types of associated disorders (Rosselli and Ardila 1996). Different types of errors can be found in calculation tasks:

(1) Errors in numbers, such as: errors in understanding the value of the numbers (inability to recognize which number is larger (eg, 96 or 102), lexicalization (127 = 10027), decomposition (137 = 13, 7), inversions (21 = 12), substitutions (hierarchy errors; eg, 500 = 50; or, order, eg, 5 = 6), omissions (eg, 712 = 71), additions (eg, 23 = 233), transcoding errors (7 = 9; 3 = eight).

(2) "Carrying" errors: carrying is omitted or inappropriately used.

(3) "Borrowing errors": difficulties with "zero"; "borrowing is omitted, or inappropriately used.

(4) Errors in the use of basic numerical principles, such as the use of zero and the multiplication tables.

(5) Errors in algorithms, such as: inability to complete a numerical operation, inappropriate use of the space, mixture of procedures (eg, adding and multiplying), wrong sequence (numerical operation is begun at the left), inappropriate algorithm (eg, when multiplying, numbers are placed as for dividing), and reasoning errors (ie, impossible results).

(6) Errors in the use of symbols, such as forgetfulness and substitutions of numerical symbols.

Calculation disturbances associated with different brain pathologies are diverse (anarithmetia, spatial acalculia, aphasic acalculia, etc.). Rehabilitation must focus on the specific deficits with which the patient presents. In cases of primary acalculia (anarithmetia) the rehabilitation program must be directed specifically to the fundamental computational defect. In cases of secondary acalculia, the therapy must be directed to the primary defect (spatial neglect, attention deficit, aphasia, etc.).

Patients with aphasic acalculia, when improving from their oral language defect, also significantly and concurrently improve in their calculation disturbance. Also patients with spatial acalculia, when improving from neglect, significantly improve in their written calculation ability.

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**References especially recommended by the author or editor for general reading.

Former authors

Monica Rosselli PhD (original author)

ICD and OMIM codes

ICD codes

ICD-9:
Acalculia: 784.69

ICD-10:
Acalculia: R48.8

Profile

Age range of presentation
Sex preponderance

male=female

Family history

none

Heredity

none

Population groups selectively affected

none selectively affected

Occupation groups selectively affected

none selectively affected

Differential diagnosis list

Dementia
traumatic brain injury

Associated disorders

Agraphia
Aphasia
Alexia
Alexia without agraphia
Finger agnosia
Neglect
Right-left disorientation

Other topics to consider

Alexia
Alexia without agraphia
Alzheimer disease
Extrasylvian aphasias
Neglect
Visual agnosias