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The mitochondrial inner membrane contains a large protein complex that functions in inner membrane organization and formation of membrane contact sites. The complex was variably named the mitochondrial contact site complex, mitochondrial inner membrane organizing system, mitochondrial organizing structure, or Mitofilin/Fcj1 complex. To facilitate future studies, we propose to unify the nomenclature and term the complex "mitochondrial contact site and cristae organizing system" and its subunits Mic10 to Mic60.

Mitochondria possess two membranes of different architecture and function (Palade, 1952; Hackenbrock, 1968). Both membranes work together for essential shared functions, such as protein import (Schatz, 1996; Neupert and Herrmann, 2007; Chacinska et al., 2009). The outer membrane harbors machinery that controls the shape of the organelle and is crucial for the communication of mitochondria with the rest of the cell. The inner membrane harbors the complexes of the respiratory chain, the F_1F_o-ATP synthase, numerous metabolite carriers, and enzymes of mitochondrial metabolism. It consists of two domains: the inner boundary membrane, which is adjacent to the outer membrane, and invaginations of different shape, termed cristae (Werner and Neupert, 1972; Frey and Mannella, 2000; Hoppins et al., 2007; Pellegrini and Scorrano, 2007; Zick et al., 2009; Davies et al., 2011). Tubular openings, termed crista junctions (Perkins et al., 1997), connect inner boundary membrane and cristae membranes (Fig. 1, A and B). Respiratory chain complexes and the F_1F_o-ATP synthase are preferentially located in the cristae membranes, whereas preprotein translocases are enriched in the inner boundary membrane (Vogel et al., 2006; Wurm and Jakobs, 2006; Davies et al., 2011). Contact sites

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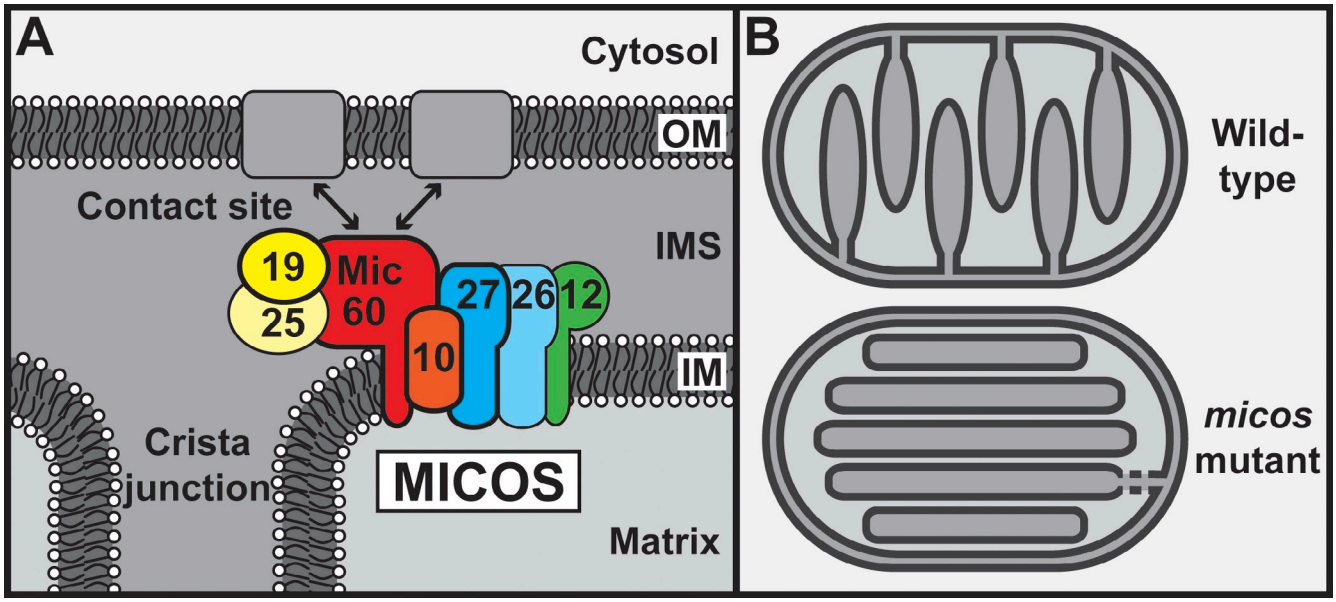

Figure 1. **MICOS complex.** (A) The MICOS complex (hypothetical model), previously also termed MINOS, MitOS, or Mitofilin/Fcj1 complex, is required for maintenance of the characteristic architecture of the mitochondrial inner membrane (IM) and forms contact sites with the outer membrane (OM). In budding yeast, six subunits of MICOS have been identified. All subunits are exposed to the intermembrane space (IMS), five are integral inner membrane proteins (Mic10, Mic12, Mic26, Mic27, and Mic60), and one is a peripheral inner membrane protein (Mic19). Mic26 is related to Mic27; however, *mic26*Δ yeast cells show considerably less severe defects of mitochondrial inner membrane architecture than *mic27*Δ cells (Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011). The MICOS complex of metazoa additionally contains Mic25, which is related to Mic19, yet subunits corresponding to Mic12 and Mic26 have not been identified so far. MICOS subunits that have been conserved in most organisms analyzed are indicated by bold boundary lines. (B, top) Wild-type architecture of the mitochondrial inner membrane with crista junctions and cristae. (bottom) This architecture is considerably altered in *micos* mutant mitochondria: most cristae membranes are detached from the inner boundary membrane and form internal membrane stacks. In some *micos* mutants (deficiency of mammalian Mic19 or Mic25), a loss of cristae membranes was observed (Darshi et al., 2011; An et al., 2012). Figure by M. Bohnert (Institute of Biochemistry and Molecular Biology, University of Freiburg, Freiburg, Germany).

between outer membrane and inner boundary membrane promote import of preproteins, metabolite channeling, lipid transport, and membrane dynamics (Frey and Mannella, 2000; Sesaki and Jensen, 2004; Hoppins et al., 2007, 2011; Neupert and Herrmann, 2007; Chacinska et al., 2009; Connerth et al., 2012; van der Laan et al., 2012).

To understand the complex architecture of mitochondria, it will be crucial to identify the molecular machineries that control the interaction between mitochondrial outer and inner membranes and the characteristic organization of the inner membrane. A convergence of independent studies led to the identification of a large heterooligomeric protein complex of the mitochondrial inner membrane conserved from yeast to humans that plays crucial roles in the maintenance of crista junctions, inner membrane architecture, and formation of contact sites to the outer membrane (Fig. 1 A). Several names were used by different research groups to describe the complex, including mitochondrial contact site (MICOS) complex, mitochondrial inner membrane organizing system (MINOS), mitochondrial organizing structure (MitOS), Mitofilin complex, or Fcj1 (formation of crista junction protein 1) complex (Table 1; Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Alkhaja et al., 2012). Mitofilin, also termed Fcj1, was the first component identified (Icho et al., 1994; Odgren et al., 1996; Gieffers et al., 1997; John et al., 2005) and was observed enriched at crista junctions (Rabl et al., 2009). Mutants of Mitofilin/Fcj1 as well as of other MICOS/MINOS/MitOS subunits show a strikingly altered inner membrane architecture. They lose crista junctions and contain large internal membrane stacks, the respiratory activity is reduced, and mitochondrial DNA nucleoids are altered (Fig. 1 B; John et al., 2005; Hess et al., 2009; Rabl et al., 2009; Mun et al., 2010; Harner et al., 2011; Head et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Alkhaja et al., 2012; Itoh et al., 2013). It has been reported that the complex interacts with a variety of outer membrane proteins, such as channel proteins and components of the protein translocases and mitochondrial fusion machines, and defects impair the biogenesis of mitochondrial proteins (Xie et al., 2007; Darshi et al., 2011; Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Alkhaja et al., 2012; An et al., 2012; Bohnert et al., 2012; Körner et al., 2012; Ott et al., 2012; Zerbes et al., 2012; Jans et al., 2013; Weber et al., 2013). The MICOS/MINOS/MitOS/Mitofilin/Fcj1 complex thus plays crucial roles in mitochondrial architecture, dynamics, and biogenesis. However, communication of results in this rapidly developing field has been complicated by several different nomenclatures used for the complex as well as for its subunits (Table 1).

To rectify this situation, all authors of this article have agreed on a new uniform nomenclature with the following guidelines. (a) The complex will be called "mitochondrial contact site and cristae organizing system" (MICOS). The protein subunits of MICOS are named Mic10 to Mic60 as listed in Table 1. (b) The names, including the numbers shown in Table 1, will be used in all organisms, e.g., Mitofilin/Fcj1 will be named Mic60 in any organism. In case the name MicX has been given to another gene/protein in an organism or a database requires a longer name, the

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Table 1. **New nomenclature of MICOS**

Standard name	Former names	Yeast ORF	References
Complex			
MICOS	MINOS, MitOS, MIB, Mitofilin complex, and Fcj1 complex		Xie et al., 2007; Rabl et al., 2009; Darshi et al., 2011; Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Alkhaja et al., 2012; An et al., 2012; Bohnert et al., 2012; Ott et al., 2012; Jans et al., 2013; Weber et al., 2013
Subunits			
Mic10	Mcs10, Mio10, Mos1, and MINOS1	YCL057C-A	Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Alkhaja et al., 2012; Itoh et al., 2013; Jans et al., 2013; Varabyova et al., 2013
Mic12	Aim5, Fmp51, and Mcs12	YBR262C	Hess et al., 2009; Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Varabyova et al., 2013
Mic19	Aim13, Mcs19, CHCH-3, CHCHD3, and MINOS3	YFR011C	Xie et al., 2007; Hess et al., 2009; Darshi et al., 2011; Head et al., 2011; Alkhaja et al., 2012; Ott et al., 2012; Jans et al., 2013; Varabyova et al., 2013
Mic25 (metazoan Mic19 homologue)	CHCHD6 and CHCM1		Xie et al., 2007; An et al., 2012
Mic26	Mcs29, Mio27, and Mos2	YGR235C	Harner et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011
Mic27	Aim37, Mcs27, APOOL, and MOMA-1	YNL100W	Hess et al., 2009; Harner et al., 2011; Head et al., 2011; Hoppins et al., 2011; von der Malsburg et al., 2011; Weber et al., 2013
Mic60	Fcj1, Aim28, Fmp13, Mitofilin, HMP, IMMT, and MINOS2	YKR016W	Icho et al., 1994; Odgren et al., 1996; Gieffers et al., 1997; John et al., 2005; Wang et al., 2008; Rabl et al., 2009; Rossi et al., 2009; Mun et al., 2010; Park et al., 2010; Körner et al., 2012; Zerbes et al., 2012; Itoh et al., 2013; Varabyova et al., 2013

APOOL, apolipoprotein O–like; HMP, heart muscle protein; IMMT, inner mitochondrial membrane protein; MIB, mitochondrial intermembrane space bridging.

name MiccX will be used in this organism, but the number will not be changed. The use of capital and small letters as well as of italics will follow species-specific conventions, e.g., in budding yeast (*Saccharomyces cerevisiae*), Mic60 will be used for the protein, and *MIC60* will be used for the gene. (c) The current names of MICOS genes and proteins in databases will be renamed according to the uniform nomenclature. This includes the names of mutants when they contain the name of a MICOS gene or protein, e.g., *fcj1*Δ mutant cells will be renamed to *mic60*Δ mutant cells. (d) In case several isoforms of a MICOS subunit are present in an organism, this will usually be indicated by -1, -2, etc. (e.g., Mic60-1 and Mic60-2 or MICC60-1 and MICC60-2). When species-specific conventions strictly require the use of A, B, or I, II, etc. for designation of isoforms, these additions will be used. (e) In case new subunits of MICOS will be identified, they will be named MicY. The number Y will be the molecular mass of the identified mature protein in kilodaltons. The same number will be used for orthologues in other organisms, i.e., these orthologues are also named MicY and thus retain the initially assigned Mic number independent of their exact molecular mass. In case a number has already been used for another Mic protein, the closest next available number will be used. The name Mic will only be given to genuine subunits of the MICOS complex, not to interaction partners or assembly factors that are not a steady-state component of the MICOS complex. (f) The names Mic14, Mic17, and Mic23 (mitochondrial intermembrane space cysteine motif proteins) that are currently used for three non-MICOS yeast proteins (Gabriel et al., 2007; Vögtle et al., 2012) will be changed to Mix14, Mix17, and Mix23 (mitochondrial intermembrane space CX_nC motif proteins) in the *Saccharomyces* Genome Database, and the new nomenclature will be used for orthologues identified in other organisms.

The MICOS complex is of central importance for the maintenance of mitochondrial inner membrane architecture and the formation of contact sites between outer and inner membranes and thus is involved in the regulation of mitochondrial dynamics, biogenesis, and inheritance. We expect that the uniform nomenclature will facilitate future studies on mitochondrial membrane architecture and dynamics.

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