Experience in the corrective treatment of patients with atrioventricular septum

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Abstract

Introduction: *Atrioventricular septal defects are a wide spectrum of cardiac malformations, from partial until complete with one unique atrioventricular valve, atrioventricular valve communication, and leaky left heart valve. Its fast evolution to pulmonary vascular disease calls for early surgical management. Corrective treatment has a high percentage of re-operations and 8.6% mortality.* **Objectives:** *To describe the results of corrective treatments of atrioventricular septum defects in our institution's patients.* **Materials and methods:** *Observational, cross-sectional, analytical, and retrospective study of the atrioventricular septum defect patients during the period from March 2013 until March 2015.* **Results:** *51 atrioventricular septum defect patients were operated, nine with incomplete defect and 42 with complete defect, predominance type A of Rastell (35, 81.3%). Age at diagnosis was from 2.9 to 7.4 months; 82.3% of the patients have Down's syndrome. The cardiopathy with more association was the patient ductus arteriosus in 61.9% of cases. Average stay in intensive care was 3.8-9.9 days. Eight (15.6%) patients died.* **Conclusion:** *Diagnosis and surgical treatment of atrioventricular septum defects in our institution it is made early. Results from surgical correction are comparable to that reported in the international literature.*

KEY WORDS: *Atrioventricular septum defect. Atrioventricular canal. Congenital heart disease.*

Introduction

Congenital heart defects are structural defects of the heart and large vessels that are produced during embryonic and fetal development[1]. They are the most common congenital malformations. Their incidence in developed countries is estimated to range between 5.2% and 12.5% of live births, and around 1% in the general population. In the USA, congenital heart defects constitute the main cause of neonatal defect-associated death, with more than 6000 lives claimed per year. In our country, their real prevalence is unknown; as a cause of children's death, it is ranked at sixth place in those younger than 1 year, an as the third cause in children aged between 1 and 4 years[2]. In Mexico, 50% of cardiac defects require surgical procedures and, out of these, 30% are carried out with cardiopulmonary bypass[3]. Eighty-five percent of heart defects are known to be regarded as multifactorial. The National Institute of Statistics, Geography and Informatics, on its 2011 report, refers a total of 29,050 deaths in children younger than 1 year, out of which 3485 died due to congenital malformations of the circulatory system[4].

Treatment of atrioventricular septal defects

Heart malformations are divided in two large groups: those characterized by the presence of cyanosis as predominant clinical feature and those that are acyanotic[5]. In the USA, in a study conducted between 1940 and 2002 by Hoffman's group, 1.2 million children were reported to have born with any heart defect

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classified as simple (ventricular septal defect [VSD], patent ductus arteriosus [PDA], atrial septal defect [ASD], mild pulmonary valve stenosis), 600,000 children with any moderate heart defect (aortic valve or pulmonary valve stenosis, non-critical coarctation of the aorta) and approximately 500,000 with any complex heart defect, including atrioventricular septal defects (AVSD)[6].

By definition, AVSD can be considered as an absence or deficiency of the AV septum as a consequence of endocardial cushions defective development between the 4th and 5th gestation weeks, which causes communication at the interventricular septum inlet portion, a common valve annulus and a common AV valve[7]; it encompasses a wide spectrum of lesions, from ostium primum (OP)-type ASD with mitral cleft to complete AV canal[8].

This heart defect accounts for about 3% of congenital heart malformations in our country[9] and for 7% in the USA[10], and owing to its complex embryonic origin, it is currently accepted that it can be divided into: a) complete defects, where an OP interatrial defect is found in continuity with the inlet interventricular defect and a mitral cleft, with a single common valve annulus with 5 or 6 leaflets or valves[7], and b) partial defects, where there is no inlet VSD, with only the atrial septal defect and the mitral cleft being present, with two subtypes, which are intermediate and transitional AVSD. Intermediate AVSD is characterized by a single AV valve annulus that is divided by a tongue of tissue into right and left orifices; transitional AVSD has two separate AV valve annuli in addition to an OP ASD and mitral cleft[11].

In addition to alterations in septal and valve structures of the heart, there are other anomalies added in the ventricular geometry, the fibrous skeleton and the conduction system[12].

This heart defect can occur as an isolated malformation, although often it is associated with other anomalies, such as left obstructive lesions (aortic coarctation, aortic arch interruption), double ventricular inlet, discordant ventriculoarterial connection and right ventricle double outlet, in addition to tetralogy of Fallot or pulmonary stenosis in approximately 10% of patients[8]. In addition, often it is part of genetic syndromes, such as Ellis-van Creveld syndrome, Mohr ectodermal dysplasia and Down syndrome[12], and it has even been considered as an exclusive heart defect of patients with trisomy 21, who represent up to 80% of all diagnosed cases[13], even considering that tetralogy of Fallot's association with AVSD occurs more frequently in patients with trisomy 21 than in those with normal karyotype[11]. In recent literature, there is only one cohort study available encompassing the time period between 1958 and 1997, reporting that patients with Down syndrome and AVSD have high mortality in postsurgical outcomes in comparison with the rest of the population without Down syndrome[14], although, currently, with an adequate surgical technique and correct postsurgical care, trisomy 21 is considered not to represent by itself a risk factor for postsurgical evolution; however, there is no study available so far where we can observe these factors.

The description by Rastelli, published in the decade of 1960 and applicable to complete AVSD, for the position of the septal leaflets in relation to the interventricular septum is still in use and is accepted when surgical options are considered for AV canal repair[15]. It is based on the bridging leaflet or left anterior leaflet site of insertion onto the interventricular septum across its papillary muscle[16], which divides complete AVSD into 3 types: in type A, the bridging leaflet is mostly contained within the left ventricle and anchored to the crest of the ventricular septum with chordae tendinae (the most common); in type B, the bridging leaflet extends towards the right ventricle and is supported by right ventricle anomalous papillary muscle emerging from the septomarginal septum; in type C, the bridging leaflet floats freely without attachments to the septum, extends further to the right ventricle and is connected to an anterior papillary muscle[8].

In addition to these structural anomalies, the heart conduction tissue is shifted towards the most posterior part of the VSD, which produces a left-deviated electrical axis on the electrocardiogram, in addition to first grade AV block and right bundle branch block[7].

Patients with AVSD are usually symptomatic within the first year of life, but pulmonary vascular disease develops within the first months, and early surgery is therefore indicated[15].

Complete AVSD typical hemodynamic lesion is the result of high volume overload produced by valvular insufficiency, VSD and OP ASD[7]. Symptoms include growth retardation, fatigue with feeding and data consistent with heart failure, such as tachycardia, tachypnea and hepatomegaly[8]. On auscultation, cardiac sounds may exhibit an increase in first heart sound intensity and fixed unfolding of the second sound, with reinforcement of the pulmonary component depending on the degree of associated hypertension[17]. The presence of murmurs is variable, and from a soft systolic

murmur to a grade IV/VI holosystolic murmur originating in the VSD can be found[8,17]. Patients who are AVSD carriers with mild mitral insufficiency usually present with a clinical picture that is similar to an isolated interatrial defect, and can evolve asymptomatically during the first years of life.

For diagnosis, in addition to physical examination findings an electrocardiogram characteristic data, an echocardiogram in expert hands can provide sufficient information for complete presurgical assessment, with heart catheterization being left for cases with associated structural anomalies or if there is suspicion of pulmonary vascular disease[15]. A thorough echocardiographic evaluation should assess the size of the atrial septal defect, completely examine the single AV valve, its insertion site, presence or not of cleft, the degree of insufficiency, the position of the bridging leaflet with regard to the ventricular septum, the size of the ventricular septum defect, ventricular size, define if it is a balanced or unbalanced canal (if the single AV binding is evenly distributed between both ventricles it is considered to be of the balanced type)[11], left ventricle outlet tract and presence or not of associated heart anomalies.

Given that patients with this heart defect progressively evolve towards pulmonary vascular disease within the first year of life, and considering that AVSD causes 27% of mortality during the first 6 months of life and 40% by the end of the first 12 months, surgical repair is thought should be carried out early, at around 4-6 months of age[10].

AVSD repair surgical outcomes have been the focus of attention of specialized centers from all over the world and have been used as a representation of congenital heart defects surgical management experience in different institutions[18].

The first AVSD correction was carried out by Lillehei in 1955, with direct suture of the OP border onto the crest of the ventricular septum. Subsequently, in 1962, the single-patch technique was introduced by Maloney[7]; in 1978, the double technique started being used[15] and, later, in 1990, Wilcox and Nunn shunted the closure with a single, modified patch, closing the VSD with loose stitches and the OP ASD with a pericardial patch[7].

Mortality of AV canal surgery was high at up to 10%[7] but, currently, thanks to advances in the management of cardiovascular anesthesia techniques, cardiopulmonary bypass, postsurgical intensive care and innovation in surgical techniques themselves, it has been able to be lowered down to 3%[19]. The surgical repair outcome depends largely on a left AV valve function recovery, the failure of which is reported in patient follow-up at immediate postoperative period in 6% to 20% with varying degrees of insufficiency[10], which is generally progressive.

With regard to the patients with Down syndrome, complete biventricular repair has been shown to entail the same benefits than in patients with chromosomal normality, without operative risk being increased; hence the importance of early diagnosis and opportune treatment, since late repair reduces survival by up to 58%[20].

Congenital heart defects are known to be the most common birth malformations. AVSD leads to pulmonary vascular disease premature development, which implies that early detection and opportune corrective treatment should be established in these patients in order to prevent the development of irreversible pulmonary disease.

In spite of advances in surgery for the correction of AVSD, there is still a high percentage of reoperations, caused in the first place by left AV valve residual insufficiency and, secondarily, by left outlet tract obstruction, interventricular residual defects or electrical conduction alterations, such as postsurgical AV block, which require pacemaker placement.

In Mexico, there are eight tertiary care specialized centers where this surgery is performed; however, there are no publications on these patients' corrective management outcome with national data.

In our hospital, during the year of 2011, heart septa congenital malformations were at second place in outpatient care, only surpassed by ischemic heart disease. Given that this is the reference center of the northeastern region, with a population of 10,168,215 affiliates covered up to the year 2010, approximately 300,000 of them will have some malformation involving the development of endocardial cushions.

In view of the above, interest arose in knowing the results obtained in the corrective management of patients with AVSD in our institution, UMAE No. 34 Cardiology Hospital.

Congenital heart defects are the main cause of noninfectious death in newborns around the world. The reported prevalence per every 1000 live births ranges from 2.17 in the USA and Canada to 8.6 in Spain and to up to 12.3 in Italy[21,22]. In our country, they are the second cause of death in children younger than 1 year since 2005, and as the third cause of death at ages between 1 and 4 years. Considering the annual birth rate in Mexico, it can be assumed

that approximately 200,000 children are born with some type of congenital heart defect every year, out of which from 3% to 7% will have an AV septal defect. In addition, considering that trisomy 21 is the most common chromosomal alteration in human beings, with an incidence of 1 for every 680 live births[20], out of which up to 50% will be carrier of some AVSD, it is important for AV septum defects to be considered as a cause of morbidity and mortality in our current population.

The purpose was to describe the experience in the corrective treatment of patients with AVSD attended to at the IMSS No. 34 UMAE Cardiology Hospital.

Specific objectives were:

- To describe demographic characteristics and associated comorbidity in patients with AVSD.
- To identify the AVSD most common type.
- To determine the type of surgical technique employed in patients with AVSD and its main complications.
- To know the mortality rate and the direct cause of death.

Material and methods

Inclusion criteria: all patients with AVSD attended to at No. 34 UMAE were included. The totality of AVSD-diagnosed patients that were brought to corrective surgery at the IMSS No 34 UMAE Cardiology Hospital within the period from March 1, 2012 to March 31, 2015 was included.

The results were assessed by means of descriptive statistics using central tendency measures and inferential statistics for categorical variables. The chi-square test was used, and for numerical variables, Student's t or Mann Whitney tests were employed. Version 20 of the SPSS statistical package was used, with data being presented in graphs and tables.

Ethical aspects

According to the General Statute of Health in Matters of Research for Health, this protocol was considered risk-free, and no informed consent letter was therefore required. In addition, the protocol was evaluated and approved by the Local Committee of Investigation, in adherence to institutional regulations and ethical principles in matters of research.

In spite of this project being retrospective and that only records of congenital heart defect patients' medical files were worked with, patient information and identity were kept under confidentiality according to considerations issued in the Nuremberg Code and the Declaration of Helsinki issued in 1964 and its various revisions, including 2013 Brazil update, as well as to international standards for medical research involving human subjects, adopted by the World Health Organization and the Council of International Organizations of Medical Sciences.

Data collection was carried out by means of medical records review, with information regarding age at diagnosis and surgical intervention, gender, type of AVSD, performed surgery, cardiopulmonary bypass time, aortic clamping time, trans-surgical complications, days of intensive care unit stay, postsurgical complications, vitality at intensive care unit discharge and cause of death being recorded. These data were captured in a previously prepared case report form and subsequently were concentrated in a database created with the Microsoft Office Excel package and were processed to reach conclusions.

Results

Fifty-one patients were intervened, out of which 23 (45%) were females and 28 (54.9%) were males. Their origin was from the following States of the Republic: Nuevo León, 21 patients (41.1%); Tamaulipas, 10 patients (19.6%); Coahuila, 6 patients (11.7%); San Luis Potosí and Chihuahua, 4 patients (7.8%) each; and Zacatecas, 3 patients (5.8%).

The type of AVSD was divided in complete in 42 patients (82.3%) and incomplete in 9 (17.6%), with the most common among complete defects according to Rastelli's classification being type A, with 35 patients (81.3%), followed by type B with 6 patients (13.9%) and type C with 1 patient (2.3%). Among incomplete defects, the most common was inlet VSD in 8 patients (88.8%), and there was only 1 patient (11.1%) with inlet VSD and OP ASD.

Mean age at diagnosis was 2.9 ± 7.4 months, with a range from 1 day of life to 4 years of age, with incomplete AVSD carriers having been diagnosed at later ages.

In the complete AVSD group, the most common associated heart defect was PDA in 26 patients (61.9%), followed by the tetralogy of Fallot in 2 patients (4.7%); other associations were right heterotaxis, supracardiac anomalous pulmonary venous connection, aortic arch interruption, ostium secundum-type ASD, vascular annulus, muscular VSD and pulmonary atresia, with

1 patient (2.3%) each. Of this group of patients, 4 (9.3%) had previous pulmonary banding surgery.

Incomplete AVSD was also more commonly accompanied by PDA in 5 patients (55.5%), and ostium secundum-type ASD and aortic arch hypoplasia, with 1 patient (11.1%) each.

The extracardiac comorbidity found in the highest number of patients was Down syndrome, which was shown by 42 patients (82.3%), followed by congenital hypothyroidism in 24 (47%), polydactyly in 3 (8%) and anorectal malformation in 2 (3.9%). Other conditions were also observed (Table 1).

At the moment of surgery, patients with complete AVSD had a mean age of 7.6 ± 6.6 months. Patients with incomplete AVSD were operated with a mean age of 15.3 ± 23.9 months.

Thirty-four corrective surgeries (81%) and 8 palliative surgeries (19%) were practiced in the group of complete AVSD patients. Among the corrective surgeries, the two-patch technique was the most commonly used, with 19 patients (55.8%), followed by the Australian technique in 14 patients (41.1%). Correction of the Fallot tetralogy was performed at the same surgical time in one patient. The palliative surgeries performed were modified Blalock-Taussig systemic-pulmonary fistula in 4 patients (75%), pulmonary artery banding in 2 patients (25%) and bidirectional cavopulmonary bypass in 1 patient (12.5%).

In the group of patients with incomplete AVSD, 100% were brought to corrective surgery with VSD closure and, in one, aortic plasty was also performed.

All corrective surgeries were carried out under cardiopulmonary bypass. Aortic clamping and cardiopulmonary bypass times in complete defects were 116.13 ± 48.9 minutes and 160 ± 72.8 minutes, respectively. In incomplete AVSDs, aortic clamping and cardiopulmonary bypass times were 43.4 ± 19.7 minutes and 61 ± 25.8 minutes, respectively.

Trans-surgical complications occurred in 7 patients (13.7%), with complete AV block and cardiopulmonary arrest being the most common, with 3 patients (42.8%) each, followed by ventricular fibrillation in 1 patient (14.2%).

In the postsurgical period, main complications were pneumonia in 10 patients (19.6%), complete AV block in 9 patients (17.6%), acute renal lesion in 8 patients (15.6%), sepsis in 4 (7.8%) and pneumothorax in 4 (7.8%); the rest of complications are shown in table 2.

Intensive care unit mean length of stay was longer for patients with complete AVSD, with 9.9 ± 3.8 days and a range between 1 and 90 days (in one patient complicated with VSD patch detachment, pneumonia and hospital-acquired sepsis). In patients with incomplete AVSD, mean intensive care unit length of stay was 6.22 ± 2.9 days.

Eight patients (15.6%) died, and the most common cause was cardiogenic shock in 6 patients (75%), followed by ventricular fibrillation in 1 patient (12.5%) and septic shock in 1 patient (12.5%).

Table 1. Comorbidity in patients with AVSD

Comorbidity	Frequency	%
Down syndrome	42	82.3
Congenital hypothyroidism	24	47
Polydactyly	3	5.8
Anorectal malformation	2	3.9
Bochdalek hernia	1	1.9
Epilepsy	1	1.9
Cholestatic syndrome	1	1.9
Laryngomalacia	1	1.9

Table 2. Postsurgical complications

Complication	Frequency	%
Pneumonia	10	19.6
Complete AV block	9	17.6
Acute renal lesion	8	15.6
Sepsis	4	7.8
Pneumothorax	4	7.8
Pleural effusion	3	5.8
Pulmonary hypertension crisis	3	5.8
Cardiopulmonary arrest	3	5.8
VSD patch detachment	1	1.9
Pericardial effusion	1	1.9
Seizures	1	1.9
First-degree AV block	1	1.9
Atelectasias	1	1.9
Multiple organ failure	1	1.9
Necrotizing enterocolitis	1	1.9
Hemothorax	1	1.9

Discussion

In this retrospective study, the results obtained with AVSD surgical correction are similar to reports in the

international literature. In the grouping by subtypes, in an anatomic-embryological analysis carried out at the Ignacio Chávez National Institute of Cardiology, the group corresponding to Rastelli's classification type A was found to occupy 44% of cases, followed by group B in 32% and subsequently group C in 24%[12], which is consistent with our results.

With regard to opportune diagnosis, it can be claimed that our institution and referral areas are carrying out an adequate screening for congenital heart defects, since diagnosis in patients with AVSD was confirmed prior to 6 months of age. This influences on early surgical management, thus decreasing complications with regard to pulmonary vascular disease development, which in this heart condition is known to occur at earlier ages, considering that at 12 months of age there are already irreversible changes[17]. In patients with incomplete AVSD, diagnosis at later ages is due to the heart defect hemodynamical behavior, where the increase in pulmonary flow only occurs in relation to the VSD size.

As for associated heart conditions, so far we only know what Díaz et al. reported[17], who refer that the tetralogy of Fallot is the most commonly found heart malformation, followed by large arteries transposition, Ebstein anomaly and right ventricle double outflow; as well as findings published by Hoohenkerk et al. in 2010[19], with a frequency of 11.5% for the tetralogy of Fallot as associated heart condition. In our review, we found PDA to be the most commonly associated heart defect (61.9%), followed by the tetralogy of Fallot. Of note, none of the patients in our series had Ebstein anomaly, large arteries transposition or right ventricle double outflow, which may suggest the necessity to carry out a review with a larger number of patients with AVSD in order to more certainly establish associations with other heart anomalies that are not mentioned in the international literature.

The results obtained for the relationship between AVSD and Down syndrome are consistent with reports in different studies, which refer that more than 40% of patients with Down syndrome have AVSD and 25% have VSD[10]. This was reflected in our patients, in whom 82% of AV canal-associated extracardiac morbidity was Down syndrome. Considering this important association, the possibility of a new investigation is opened in order to determine if patients with Down syndrome and AVSD brought to surgical correction have higher mortality in comparison with the population with normal karyotype, as referred by Patel et al.[23] on their report published in 2012.

Surgical correction in our health institution was practiced in 81% of patients; out of these, 80% was within the age range between 10 days and 12 months, with a higher prevalence between 30 days and 6 months of age, which is consistent with findings reported by Tumanyan et al.[10] in their case series, with an age range of 5.6 ± 3 months for surgically intervened patients.

All corrective surgeries were performed with cardiopulmonary bypass and aortic clamping, with times of 55 to 230 minutes for bypass and 71 to 360 minutes for aortic clamping. Comparing these figures with those reported by Kaza et al.[18], who refer clamping times between 31 and 82 minutes and bypass times between 36 and 112 minutes, we consider that, in spite of the differences, these times are not a determining factor for the rate of complications or mortality in our patients. Surgical techniques used in our hospital center were equally applied in the series by Weintraub et al.[24] and Suzuki et al.[25], who refer that, currently, the two-patch technique is the most widely used in specialized centers for the management of congenital heart defects; in addition, in continuing with the update in the management of this heart defect, the Australian technique has been progressively substituting other techniques, owing to a lower frequency of complications and its rapidness[26].

Complications reported as secondary to AVSD trans-surgical and post-surgical management encompass a wide variety, with the most common being those associated with infectious processes, arrhythmias and pulmonary hypertension crises; between 10% and 20% of cardiosurgery-operated patients are known to develop hospital-acquired infections[27]. Hoohenkerk et al.[19] report heart failure as the main complication in their series of AVSD postoperated patients with 11%, followed by sepsis with 6% and gastrointestinal bleeding, whereas Tumanyan et al.[10], in their study series, refer pulmonary hypertensive crisis as the most common complication in 16% of cases, followed by infectious complications in 29%. In our series, the most commonly found complication was pneumonia in 19.6% of cases, followed by arrhythmias (complete AV block) in 17.6% and acute renal lesion in 15.6%, which is consistent with findings reported in the aforementioned studies and with Brown et al. reports[28], which refer 6.5% of acute renal lesion incidence in 342 open heart surgery post-operated patients. With regard to the infectious processes etiology, only in two patients were the specific

microorganisms isolated, with these being *Staphylococcus aureus* and *Stenotrophomonas maltophilia*. In the series of patients published by Tumanyan et al.[10] in 2015, 16 out a total of 152 patients required definitive pacemaker implantation owing to AV block at early postoperative period; in our study, only two patients required definitive pacemaker implantation, and the rest of those who had AV conduction block, were managed with steroids at anti-inflammatory doses, with remission of the block being achieved.

Of note, in contrast with reports in different international publications with regard to right AV valve insufficiency as a cause for surgical reintervention at postsurgical short-term period[18,19], no patient was reported to require reoperation for this reason in our study. In patients who had right AV valve insufficiency documented in postsurgical echocardiographic control, it was of mild grades and responded adequately to pharmacological management with diuretics.

Kaza et al.[18], in their study published in 2011, refer that postsurgical patients' intensive care unit mean length of stay is 9.9 days; this is probably due to the prolonged 90-day stay of a single patient whose postsurgical evolution got complicated.

With the emergence of new surgical techniques, which in turn enable higher experience in AVSD repair, the results have gotten to be regarded as excellent, with a progressively decreasing mortality[29]. Currently, the outcomes in the surgical management of this heart defect are one of the indicators used to determine the experience of the surgeon and of the specialized health institution[30,31], and knowing the mortality of these patients at each center that treats congenital heart defects therefore allows for a referendum oriented to improve comprehensive care of patients with heart conditions to be established and, hence, to improve their quality of life and survival prognosis. Mortality in our health institution was 15.6%, with causes being cardiogenic shock, ventricular fibrillation and septic shock. If this percentage is compared with the results described by Hoohenkerk et al[19], with a mortality of 8.3% in their series, and by Studer et al.[32] and Tumanyan et al.[10], with a mortality between 14% and 16%, our results in the surgical management of these patients can be regarded as being within acceptable ranges.

Conclusions

The outcomes in surgical management of patients with AVSD at our institution are within the parameters referred in different comparative trials from the rest of the world.

Currently, in our hospital center there is a marked tendency to first of all correct these septal defects, leaving the use of palliative surgeries only for those patients with a heart anatomy that entails more risk than benefit when performing primary correction; in the same way, the surgical team has a tendency towards the use of new surgical techniques that decrease the risk of surgery-inherent complications and also reduce the time of cardiac arrest, which benefits myocardial fibers recovery and consequent patient survival improvement.

Up to this moment, adequate screening for congenital heart defects is carried out in the northeastern region of our country, which has enabled early diagnosis and surgical correction at ages where irreversible pulmonary vascular disease has not yet developed, which considerably influences on improvement of these patients' prognosis.

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