

Truncus arteriosus communis with intact ventricular septum¹

IAN CARR, SAROJA BHARATI, V. S. KUSNOOR, AND MAURICE LEV

From the Divisions of Pediatric Cardiology and Pathology, Cook County Children's Hospital, Chicago, Illinois; and The Congenital Heart Disease Research and Training Center, Hektoen Institute for Medical Research, Chicago, Illinois, USA

SUMMARY This is the first documented case of truncus arteriosus communis with intact ventricular septum in which extensive clinical, haemodynamic, angiographic, and pathological data are available. Angiography suggested the presence of two discrete semilunar valves but necropsy showed a basically single semilunar valve.

This case fills a gap in the spectrum of aorticopulmonary, truncal, and infundibular septal defects, and reinforces the belief that the essence of truncus arteriosus communis is a single semilunar valve, common to both ventricles, which need not be associated with the defects in the adjacent parts of the structural continuum.

In the common forms of truncus arteriosus communis, as the term is used in cardiology, a single great artery arises from the heart above a single semilunar valve, gives origin in its ascending portion to coronary and pulmonary arteries, and continues as the aorta. Though an infundibular septal deficiency is not a necessary component of truncus arteriosus communis, instances of this anomaly in which the infundibular septum is intact are very rare. We are aware of no previous report describing a heart with truncus arteriosus communis and intact ventricular septum in which extensive clinical, haemodynamic, and angiographic data, as well as pathological data are available. The purpose of this report is both to describe and to discuss such a case and to comment on its implications with respect to the embryology, definition, and classification of truncus arteriosus communis.

Case history

A 3-month-old boy had been born at term by caesarean section, to a diabetic mother; his birth-weight was 4252 g (9 lb 6 oz). On the second day of extrauterine life he was noticed to have a heart murmur. Mild central cyanosis and congestive

heart failure were first detected at the age of 3 weeks. He had frequent respiratory infections and persistent congestive heart failure in spite of full medical treatment.

PHYSICAL EXAMINATION AT AGE 3 MONTHS

The infant was in mild respiratory distress with a respiratory rate of 60/minute. There was slight central cyanosis and no clubbing. Pulse rate was 160/minute, regular in rhythm, and bounding in form. Femoral pulses were neither diminished nor delayed. There was mild intercostal retraction on inspiration, and easily visible cardiac pulsation on the left side. The apex beat was in the 5th left interspace beyond the mid-clavicular line. There was a slapping parasternal impulse and a systolic thrill maximal in the 3rd and 4th spaces at the left sternal edge. On auscultation the first sound complex was of normal intensity. The second sound appeared to be split but a phonocardiogram was not obtained. A third heart sound was heard at the apex. A grade 4/6 harsh systolic murmur was maximal in the 3rd and 4th spaces at the left sternal edge. A grade 2/4 mid-diastolic murmur was heard at the apex. The lungs were clear. The liver was palpable 2 cm below the right costal margin.

CHEST x-RAY

The cardiomedial shadow was moderately enlarged and the pulmonary vascularity was increased.

¹This study was supported by a grant from the Children's Heart Research Foundation and by grants from the National Institutes of Health, Bethesda, Maryland, USA.

ELECTROCARDIOGRAM

This showed regular rhythm with a mean frontal plane QRS axis of $+150^\circ$. The patterns of left atrial enlargement and combined ventricular hypertrophy were present (Fig. 1).

CARDIAC CATHETERISATION

Catheterisation at the age of 3 months (Table) showed that neither the aorta nor the pulmonary artery was entered from the right ventricle.

ANGIOCARDIOGRAMS

Injection into the root of the great artery arising

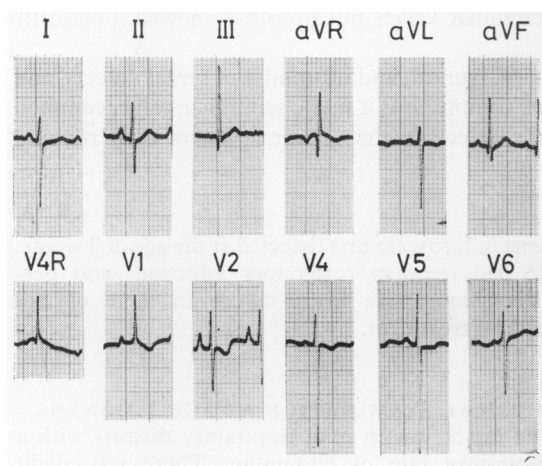


Fig. 1 *Electrocardiogram at age 3 months.*

Table *Cardiac catheterisation data at age 3 months*

Site	O ₂ saturation (%)	Pressure (mmHg)
Superior vena cava	54	
Right atrium, high	52	
Right atrium, middle	58	a = 10 mean = 7
Right atrium, low	68	
Inferior vena cava	62	
Right ventricle	68	124/0-1
Right upper pulmonary vein	95	
Left pulmonary vein	94	
Left atrium	91	a = 15, v = 25 mean = 17
Left ventricle	89	110/0
Aorta	81	mean = 52

Note: Difference in right ventricular and left ventricular systolic pressures is consistent with lack of interventricular communication and some degree of right ventricular infundibular stenosis. Aortic pressure was overdamped. The left atrial oxygen saturation, lower than that in the pulmonary veins that were entered, is presumably caused by the perfusion of under-ventilated lung whose pulmonary vein was not entered. The same explanation may apply to the left ventricular saturation with the additional factor of semilunar valve regurgitation.

from the base of the heart showed that it gave rise to the coronary, pulmonary, and systemic arteries in its ascending portion. An unopacified area at the root of this great artery, best seen in the lateral projection, was interpreted as a septum between 2 semilunar valves (Fig. 2 and 3). No reflux into either ventricle was seen.

A left ventriculogram showed opacification of the root of the great artery but not of the right ventricle.

A right ventriculogram showed opacification of the root of the great artery via a well-defined but

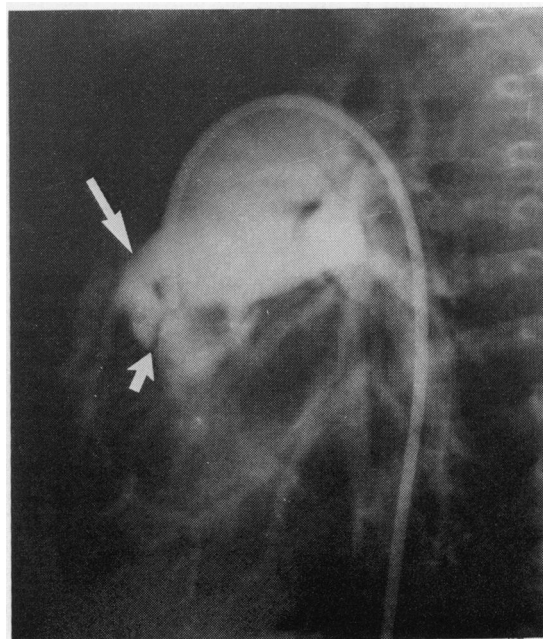


Fig. 2 *Angiogram in root of great artery showing filling defect caused by dysplastic truncal valve tissue of anterior leaflet (shorter arrow). Longer arrow indicates anterior sinus of Valsalva.*

narrow infundibulum. Opacification of the left ventricle was seen in this injection.

The diagnosis of an unusual form of truncus arteriosus, or aorticopulmonary septal defect with 2 semilunar valves, was made on the basis of these data.

SUBSEQUENT COURSE

The infant continued in severe congestive heart failure with poor weight gain, and at the age of 5 months was submitted to operation. It was planned to attempt construction of separate aortic and pul-

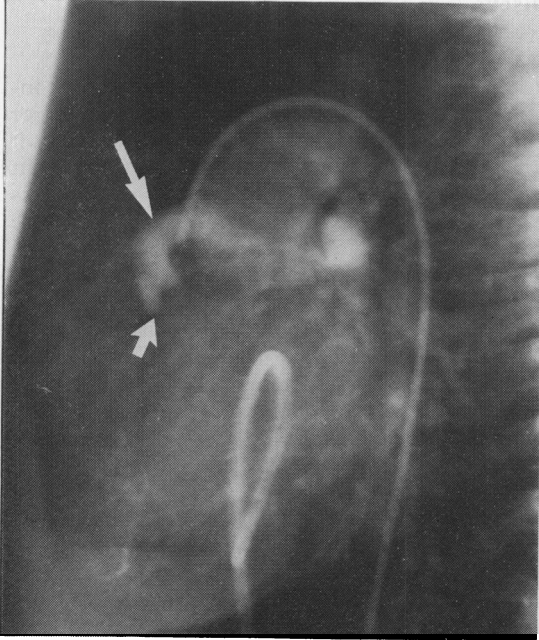


Fig. 3 Angiogram in root of great artery showing filling defect caused by dysplastic truncal valve tissue of anterior leaflet (shorter arrow). Contrast medium is trapped in anterior sinus of Valsalva (longer arrow) giving the false impression of a rudimentary main pulmonary artery with its own semilunar valve.

monary channels or, if this proved to be impracticable, to band the pulmonary arteries. During induction of anaesthesia he developed cardio-respiratory arrest. All resuscitative measures were unsuccessful.

NECROPSY FINDINGS

The heart was enlarged. From the base of the heart a single great artery emerged. There was hypertrophy, enlargement, and endocardial thickening of all the cardiac chambers. A patent foramen ovale was present.

The architecture of the infundibulum of the right ventricle was abnormal. The septal band group proceeded to the base and met the parietal band to form an arch. This arch, however, was somewhat narrowed superiorly. This was related to the presence of the single great artery emerging from both ventricles, straddling the ventricular septum. The valve of this artery was quadricuspid, with one each of anterior, posterior, right, and left leaflets. The right was slightly larger than the posterior leaflet which in turn was larger than the left; the anterior leaflet was the smallest. The commissure of the left and anterior leaflets was also at the anterior,

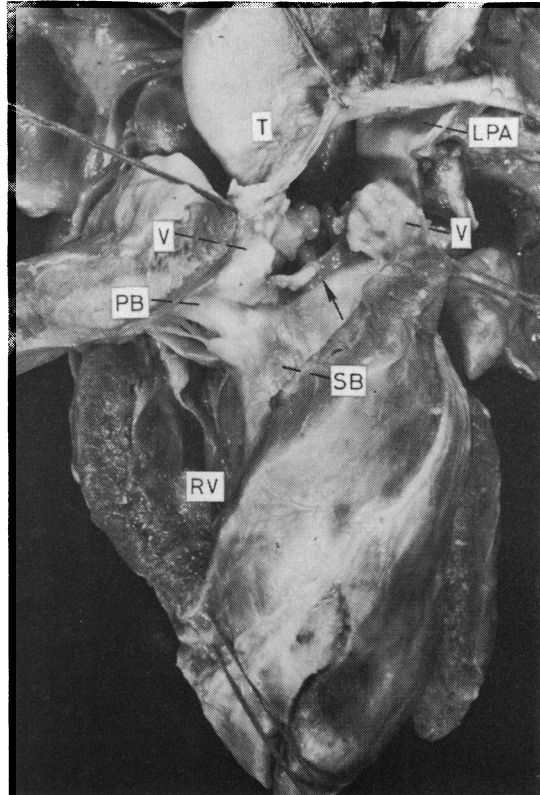


Fig. 4 Anterior view of heart showing the outflow tract of the right ventricle. RV, right ventricle; PB, parietal band; SB, septal band; V, truncal valve; T, common arterial trunk; LPA, left pulmonary artery. Arrow points to the truncal valve tissue extending over the ventricular septum. Note the absence of a ventricular septal defect.

right, side of the ventricular septum so that the left leaflet also bridged the ventricular septum. The crest of the muscular ventricular septum lay in the same plane as the arterial valve (Fig. 4 and 5). Valve tissue arose from this crest and was confluent, with the commissure of the right and posterior leaflets, posteriorly, and with the left leaflet, near its commissure with the anterior leaflet, anteriorly. The left leaflet close to its junction with the posterior, non-coronary, cusp was related to the mitral valve. The valve tissue was very thickened throughout, with many single and confluent nodular formations. The right coronary ostium emerged from the sinus of Valsalva of the right bridging cusp and the left coronary ostium from that of the left bridging cusp. These ostia appeared obstructed by the much thickened nodular formation of the valve tissue. This arterial trunk

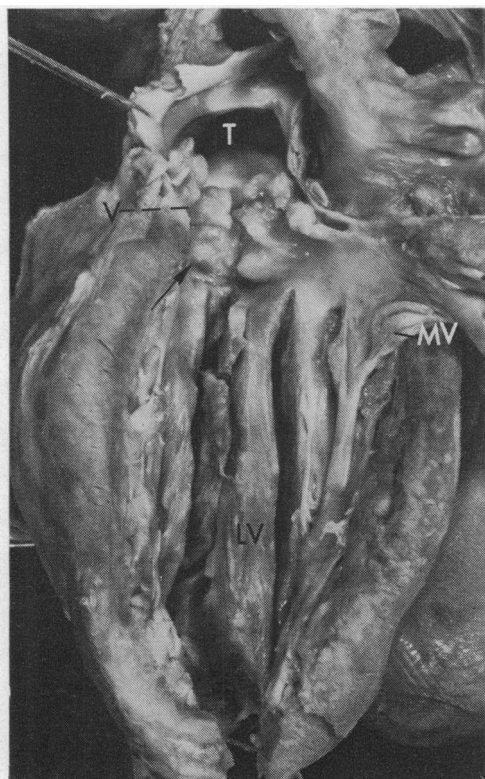


Fig. 5 Left ventricular view: the linear cuts in the left ventricle indicating the areas from which microscopical sections were studied. LV, left ventricle; MV, mitral valve; V, truncal valve; T, common arterial trunk. Arrow points to much thickened nodose truncal valve which extends on the summit of the ventricular septum. Note the absence of a ventricular septal defect from the left ventricular view.

gave off the pulmonary trunk anteriorly, which in turn divided into right and left pulmonary arteries. The main arterial trunk then proceeded into a left aortic arch giving off the brachiocephalic vessels.

The left coronary artery gave off 2 anterior descending branches but formed no left circumflex artery. The right coronary artery formed the posterior descending branch.

At a gross level all walls of the left ventricle on section presented a variegated picture of red and white areas. Microscopical sections through all walls of this chamber presented areas of recent and old infarction.

Aside from the findings in the heart the pathological diagnoses were partial pulmonary atelectasis, bilateral, with chronic passive hyperaemia of the lungs and liver.

Discussion

PREOPERATIVE DIAGNOSIS

In the angiocardiograms, the unusual diastolic configuration of the dysplastic tissue of the anterior truncal leaflet gave the false impression that it represented a septum separating an anterior right ventricular outlet valve from a posterior left ventricular outlet valve. Two semilunar valves were therefore believed to be present. Necropsy showed only one semilunar valve though the dysplastic tissue on the crest of the ventricular septum suggests an early stage of division into 2 semilunar valves. To avoid this angiographic misinterpretation in the future we need to remember this patient, and to recall how thick and fleshy the dysplastic valve tissue can be in truncus arteriosus communis.

SURGERY

Surgically, it seems that this heart may have been accessible to a procedure similar to that described by Daily *et al.* (1975). An eccentrically placed channel might have been created from the right ventricular outlet to the main pulmonary artery orifice without obstructing the channel from the left ventricular outlet to the aortic arch. The small size of the structures and the doubtful effectiveness of the dysplastic valve tissue at the right ventricular and left ventricular outlets after such a procedure would have made its success a considerable tour de force.

PATHOLOGY

The old and recent infarcts have not, to our knowledge, been previously described as complications of truncus arteriosus communis and may be related to the proximity of this valve tissue to the coronary orifices.

EMBRYOLOGY, DEFINITION, AND CLASSIFICATION

The remarkable thing about this case is the absence of an infundibular septal defect. In addition, though there is a clearly differentiated truncal valve with 4 circumferential leaflets, valvular tissue is also present on the crest of the septum extending across the diameter of a root of the great artery; this tissue is not formed into leaflets. Similar cases have previously been described by Bharati *et al.* (1974) and Rosenquist *et al.* (1976). We believe these features to have possible implications with respect to the embryology, definition, and classification of truncus arteriosus communis.

The definition and classification of truncus arteriosus communis are still the subject of considerable controversy. On the one hand there is the natural desire to relate postnatal appearances to

prenatal development, as evidenced by the use of embryological terms to describe postnatal structures, and on the other hand the pathological embryology of these defects is not known.

The normal embryonic structures whose maldevelopment is believed to be relevant to truncus arteriosus communis are the aorticopulmonary septum, the truncus arteriosus, and that part of the partition between the right ventricular and left ventricular outflow tracts immediately below the 2 semilunar valves, which becomes the infundibular septum. The term 'truncus arteriosus communis', as used by most cardiologists, includes both the single great artery arising from the heart and the semilunar valve at the root of this great artery. Some embryologists (Van Mierop *et al.*, 1978) use the term only for the precursor of 'the truncal sinuses of Valsalva and valve cusps, not the common arterial trunk distal to it'; what cardiologists call the common arterial trunk is derived from the aortic sac. The ascending aorta and pulmonary artery are formed from the aortic sac when it is partitioned by an invagination of the posterior wall called the aorticopulmonary septum. This implies that defective partition into aorta and pulmonary artery would represent, embryologically, an aorticopulmonary septal defect which may or may not be associated with persistence of the embryological truncus arteriosus communis. This also means that the term aorticopulmonary septum as used by cardiologists corresponds to the embryological aorticopulmonary septum but not the embryological truncal septum. We believe a distinction has to be made between the term truncus arteriosus communis as used to describe a prenatal, embryological structure, and truncus arteriosus communis as used to describe a postnatal structure.

If the above 3 artificial subdivisions of the structural continuum always functioned, in abnormal states, as a single developmental unit, we would not need to draw any fine distinctions between one part and another for the purposes of definition and classification. However, they do not always function as a single developmental unit in abnormal states, as evidenced, for example, by our case in which infundibular septation is apparently complete, but embryonic truncal septation is incomplete.

Fig. 6 represents a spectrum of hearts previously reported to have defects in this region, including the present heart. Though this artificially arranged sequence may not correspond completely to an embryological sequence, it may clarify the problem of definition and classification of truncus arteriosus communis for the clinician and surgeon. At one end of this spectrum (Fig. 6.1) lies truncus arteriosus

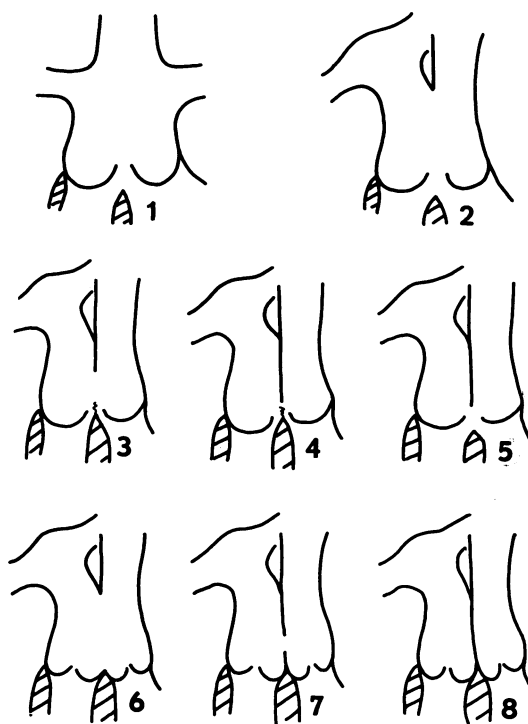


Fig. 6 Diagrammatic representation of spectrum of hearts reported to have defects in the region of the truncus arteriosus. See text. The pulmonary artery is drawn on the right side of the aorta only because of the constraints imposed by representing 3D in 2D.

communis (cardiologists' term) with no main pulmonary artery (left and right pulmonary arteries arise separately), a single semilunar valve, and failure of the infundibular septum to reach the plane of the semilunar valve, leaving what is effectively a subvalvar ventricular septal defect. This corresponds to Collett and Edwards Type 3 (Collett and Edwards, 1949) and may be regarded, embryologically, as truncus arteriosus communis with associated aorticopulmonary and infundibular septal defects. In Fig. 6.2 arterial septation has resulted in the main pulmonary artery but is otherwise the same as in Fig. 6.1. Embryologically we regard Fig. 6.2 as depicting truncus arteriosus communis with an infundibular septal defect and partial absence of the aorticopulmonary septum. The heart reported here (Fig. 6.3) exemplifies a further degree of truncus arteriosus communis (cardiologists' term) in which the infundibular septum is fully formed, there is no ventricular septal defect, truncal valve tissue is present on the crest of the infundibular

septum, but differentiation into right ventricular and left ventricular outlet valves has not occurred. Embryologically this is truncus arteriosus communis with partial absence of aorticopulmonary septum. Fig. 6.4 represents 'pure' truncus arteriosus communis with well-formed aorticopulmonary septum, embryologically, as exemplified by case 4 of Rosenquist *et al.* (1976) and Fig. 6.5 truncus arteriosus communis with no aorticopulmonary septal defect, embryologically, but with ventricular septal defect, as exemplified by case 1 of Rosenquist *et al.* (1976). The hearts described by Bain and Parkinson (1943), Van Praagh and Van Praagh (1965) and Daily *et al.* (1975) are represented by Fig. 6.6. This seems to correspond to another degree of defect in which there is little aorticopulmonary septation embryologically but separate right and left ventricular outlet valves have developed and there is no ventricular septal defect. The fact that 2 semilunar valves are present precludes the idea that there is persistence of the truncus arteriosus communis, as it is understood by embryologists. Until contrary data are available, it seems preferable to us to regard this kind of heart as an example of aorticopulmonary septal defect both embryologically and anatomically. The strictly arterial portion of these hearts is indistinguishable to us from those represented by Fig. 6.2 and 6.3. Finally (Fig. 6.7), aorticopulmonary septation progresses to the point at which the abnormality would clearly be regarded as an aortopulmonary window by the cardiologist and would correspond to an aorticopulmonary septal defect embryologically. Fig. 6.8 represents the normal.

It seems that though most instances of truncus arteriosus communis have both infundibular and aorticopulmonary septal defects as part of the complex from the embryological standpoint, suggesting a high degree of interdependence between these artificial subdivisions of the structural continuum, in rare instances, as exemplified both by this and other reported cases, one or both of these associated defects may be absent.

We believe that there are sound reasons, both embryological and surgical, for regarding aortopulmonary window, or any other instances of incomplete differentiation into ascending aorta and

pulmonary artery, with 2 semilunar valves, as representing aorticopulmonary septal defect rather than a variant of truncus arteriosus communis. Edwards (1976) and Crupi *et al.* (1977) have reached the same conclusion, but whereas they regard a single semilunar valve as an essential part of the truncus arteriosus we prefer the perspective of Van Mierop *et al.* (1978) that the single semilunar valve is the *only* essential for truncus arteriosus.

We are grateful for the assistance of Drs Gertrude Novak, Avinash Mondkar, and Constantine Tatooles.

References

- Bain, C. W. C., and Parkinson, J. (1943). Common aortopulmonary trunk: a rare congenital defect. *British Heart Journal*, **5**, 97-100.
- Bharati, S., McAllister, H. A., Rosenquist, G. C., Milles, R. A., Tatooles, C. J., and Lev, M., (1974). The surgical anatomy of truncus arteriosus communis. *Journal of Thoracic and Cardiovascular Surgery*, **67**, 501-510.
- Collett, R. W., and Edwards, J. E. (1949). Persistent truncus arteriosus: a classification according to anatomic types. *Surgical Clinics of North America*, **29**, 1245.
- Crupi, G., Macartney, F. J., and Anderson, R. H. (1977). Persistent truncus arteriosus. *American Journal of Cardiology*, **40**, 569-578.
- Daily, P. O., Sissman, N. J., Lipton, M. J., and Shumway, N. E. (1975). Correction of absence of the aortopulmonary septum by creation of concentric great vessels. *Annals of Thoracic Surgery*, **19**, 180-190.
- Edwards, J. E. (1976). Persistent truncus arteriosus: a comment. *American Heart Journal*, **92**, 1-2.
- Rosenquist, G. C., Bharati, S., McAllister, H. A., and Lev, M. (1976). Truncus arteriosus communis: truncal valve anomalies associated with small conal or truncal septal defects. *American Journal of Cardiology*, **37**, 410-412.
- Van Mierop, L. H. S., Patterson, D. F., and Schnarr, W. R. (1978). Pathogenesis of persistent truncus arteriosus. *American Journal of Cardiology*, **41**, 755-762.
- Van Praagh, R., and Van Praagh, S. (1965). The anatomy of common aorticopulmonary trunk (truncus arteriosus communis) and its embryologic implications. A study of 57 necropsy cases. *American Journal of Cardiology*, **16**, 406-426.

Requests for reprints to Dr Ian Carr, Division of Pediatric Cardiology, Cook County Hospital, 1825 West Harrison Street, Chicago, Illinois 60612, USA.